

Lipomas of cervical area – clinical and pathological considerations

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Abstract

Introduction: Cervical lipomas are tumors that may present as painless masses with slow growth. This type of tumors is usually asymptomatic until they reach a large size. They can be solitary or multiple. When multiple and symmetrical distributed around neck area, the lesions can be classified as Madelung's disease, a rare condition that affects mostly Mediterranean men. Our objective was to describe the symptoms, cytological and histological aspects, diagnostic problems and treatment strategies of the large size lipomas and to highlights that the differential diagnosis between common lipoma of cervical area and Madelung's disease can be established only on clinical grounds, with major implications in the treatment options. **Materials and Methods:** The study was retrospective and analyzed archived material. We selected four cases of cervical lipomas, each with its particularities, treated in the Clinic of Oral and Maxillofacial Surgery, Timișoara City Hospital, Romania. For all the cases, we performed a fine-needle aspiration with consequent cytological exam. The microscopic evaluation of Papanicolaou-stained slides established the diagnosis of lipomas. The surgically removed specimens were sent to the Department of Pathology. The excised specimens were prepared with routine histological technique. **Results:** The cases presented one or more lateral cervical swelling of the neck, inserted among major vascular-nervous elements, becoming life-threatening conditions. All the cases studied had the same cytological and histopathological features consistent with lipoma, fibrolipoma or angiolipoma, but, instead of this, clinical aspects differed from case to case. On clinical grounds, two patients were considered as Madelung's diseases. **Conclusions:** Despite of the same histopathological aspects, lateral cervical lipomas can be classified in varies categories with different treatment strategies and prognostic factors. The correct diagnosis of Madelung's disease is important in order to add other treatment options to the surgery or liposuction as avoiding alcohol or medication for metabolic disorders.

Keywords: lipomas of cervical area, Madelung's disease, cytology, mature adipose cell tumor.

Introduction

Cervical lipomas are solitary or multiple tumors that may present as painless masses with slow growth. The tumors are usually asymptomatic until they reach a large size [1].

When multiple and symmetrical distributed around neck area, the lesions can be classified as Madelung's disease. Madelung's disease, a rare condition also known as benign symmetric lipomatosis, multiple symmetric lipomatosis, Launois–Bensaude syndrome, is characterized by the presence of multiple, non-encapsulated fat masses in the face, neck, and other areas. Madelung's disease affects males more frequently than women with a male to female ratio of 30 to 1, having an incidence of 1 to 25 000. Even it is known as Madelung's disease, Benjamin Brodie was the first who described the condition in "St. George" Hospital, in London, in 1846, and subsequently by Otto Madelung, in 1888. Launois and Bensaude are those that termed the disease, in 1898, as benign symmetric lipomatosis [2, 3].

Madelung's disease is considered a fat metabolism

disorder. Patients with Madelung's disease present symmetric deposits of adipose tissue, usually non-encapsulated around the neck and shoulder areas. Even if it was noticed a strong correlation with alcohol abuse, the etiopathogenesis of the disease is still unknown. Moreover, Madelung's disease was diagnosed in people who do not drink alcohol. Some authors claim that abstinence from alcohol prevents progression of the disease [4, 5].

In 2002, Enzi *et al.* [6] described two types of Madelung's disease. In type I, the fat tissue is disposed as a collar around the neck and deltoid areas, while in type II the fat is diffusely accumulated as in obesity.

The diagnosis is based on clinical examination that highlights the distribution of fat masses, which do not decrease even if the caloric intake is reduced. Massive symmetric fat deposits interest parotid regions ("hamster cheeks"), neck regions ("horse's neck") and cervicodorsal regions ("buffalo hump") [1].

Our objective was to assess the symptoms, cytological and histological aspects, diagnostic problems and treatment strategies of the large size lipomas with the emphasis on

the differential diagnosis between common lipoma of cervical area and Madelung's disease, the correct diagnosis having important implications in the treatment options.

☐ Patients, Materials and Methods

The study was retrospective and analyzed archived material. We selected four cases of cervical lipomas, each with its particularities, treated in the Clinic of Oral and Maxillofacial Surgery, Timișoara City Hospital, Romania.

For all the cases, were noted the distribution, number and size of the tumors and their relationship with major nervous and vascular plexus, the symptoms that addressed the patient to the physician, the time of the evolution, life style and behavior, personal and collateral medical history. Also, were noted the signs revealed at visual inspection and palpation of the tumors and imagistic data (radiography, computed tomography). The picture was completed with the results at different laboratory tests (complete blood count, liver tests, lipid profile, glucose level, kidney function, thyroid, adrenocorticotrophic, growth and sexual hormones, cortisol with overnight suppression test and insulin).

In all the cases, preoperative cytological diagnosis was established. Fine-needle aspiration was done under local anesthesia (2% Lidocaine hydrochloride, 20 mg/mL) and palpation control. From the patients with multiple separated tumors, all the lesions were sampled. For each lesion, several passes were done in order to obtain enough cellular content. The cells were withdrawn with a syringe and spread onto microscopic slides, fixed in ethanol and stained with Papanicolaou technique. For each lesion, three slides were performed.

If the cytological diagnosis confirmed the presence of the benign tumor, in all cases the treatment consisted in surgical removal of the fatty deposits under general anesthesia (endotracheal intubation or intravenous sedation). The specimens were fixed in 10% (v/v) buffered formalin and sent to the Department of Pathology, being processed using the routine histological technique in order to obtain paraffin blocks.

For all cases, 3-μm thick sections were cut on a semi-automated Rotary Microtome Leica RM2245, mounted on histological slides and stained with Hematoxylin and Eosin (HE).

Histopathological evaluation was performed with Leica DM750 microscope.

Images were acquired using Leica DM Share system.

☐ Results

All the patients presented one or more lateral cervical swelling of the neck, inserted among major vascular-nervous elements, becoming life-threatening conditions.

Case No. 1

Clinical data

A 57-year-old man presented with a growth of soft tissues in the left anterior cervical perimandibular region (Figure 1a), with an evolution in the last four years. He was an occasional alcohol drinker and no smoker. He

addressed to the physicians for aesthetic reason. No personal or collateral medical history significant for the case was noted.

On palpation, the tumor was 4/3/3 cm in size, painless, and had soft consistency. The tumor was barely visible in the lower left oral cavity vestibule, having the appearance of an externalized odontogenic radicular cyst (Figure 1b).

Imagistic investigations

Panoramic radiography showed no dental or bone cystic changes in the target area.

Laboratory tests

The laboratory tests were without significant changes, with triglyceride, glucose and thyroid-stimulating hormone (TSH) level in normal range.

Cytological examination

The cytological examination of the aspired cells revealed that a monotonous population of rounded cells, with clear cytoplasm and eccentric located nuclei (Figure 1c), composed the tumors.

The diagnosis of presumption after cytological examination was lipoma.

Intraoperative aspects

Surgery consisted in one stage removal of the fatty mass from the left cervical and perimandibular region and it was conducted under intravenous sedation completed with local anesthesia. A horizontal skin incision, 2 cm below the lower border of the mandible was performed. After dissection, the tumor was reached and excised en bloc. A connective tissue capsule surrounding the lesion was found. After removal of the lesion, local hemostasis was done and the wound was closed in layers (Figure 1, d–f). The lesion was sent for microscopic examination.

Histopathological examination

The macroscopic evaluation of surgical specimen revealed an encapsulated yellowish tumor. On HE-stained slide, adipose tissue with fine connective septa was observed, sustaining the diagnosis of lipoma (Figure 1, g and h).

Case No. 2

Clinical data

A 55-year-old woman presented with a massive thickening of the left anterior cervical region. The increase in the neck volume was observed for the last five years, with slow progress. She presented to the physicians for aesthetic reason. She was a low drinker of alcohol and no smoker. She described no relevant past personal or collateral medical history. The physical examination revealed an enlargement of the left submandibular and upper anterior cervical areas (Figure 2, a and b). On palpation, the tumor was painless, with soft consistency and had 5/5/4 cm in size.

Imagistic investigations

A facial and cervical computed tomography (CT) scan showed a fatty accumulation in the referred areas.

Laboratory tests

The laboratory tests were without significant changes, with triglyceride, glucose and TSH level in normal range.

Cytological examination

On Papanicolaou stained smears, the mature adipose cells were admixed with spindle cells with scant cytoplasm and long, fine cytoplasmic prolongations, which led to the diagnosis of benign fibroadipose tissue (Figure 2c).

The diagnosis of presumption after cytological examination was fibrolipoma.

Intraoperative aspects

Surgery consisted in one stage removal of the tumor under general anesthesia (endotracheal intubation). A

horizontal skin incision, 4 cm below the lower border of the mandible was performed. The underlying subcutaneous tissues were carefully dissected and the tumor was exposed and removed en bloc (Figure 2, d–g). Local hemostasis was done and the wound was closed in layers. The lesion was sent for microscopic examination.

Histopathological examination

On gross grounds, the tumor was encapsulated by a thin capsule and composed of translucent areas separated by fibrous septa.

The microscopic evaluation revealed lobules of mature adipocytes delimited by thick connective tissue with hyperemiated blood vessels (Figure 2, h and i), sustaining the diagnosis of fibrolipoma.

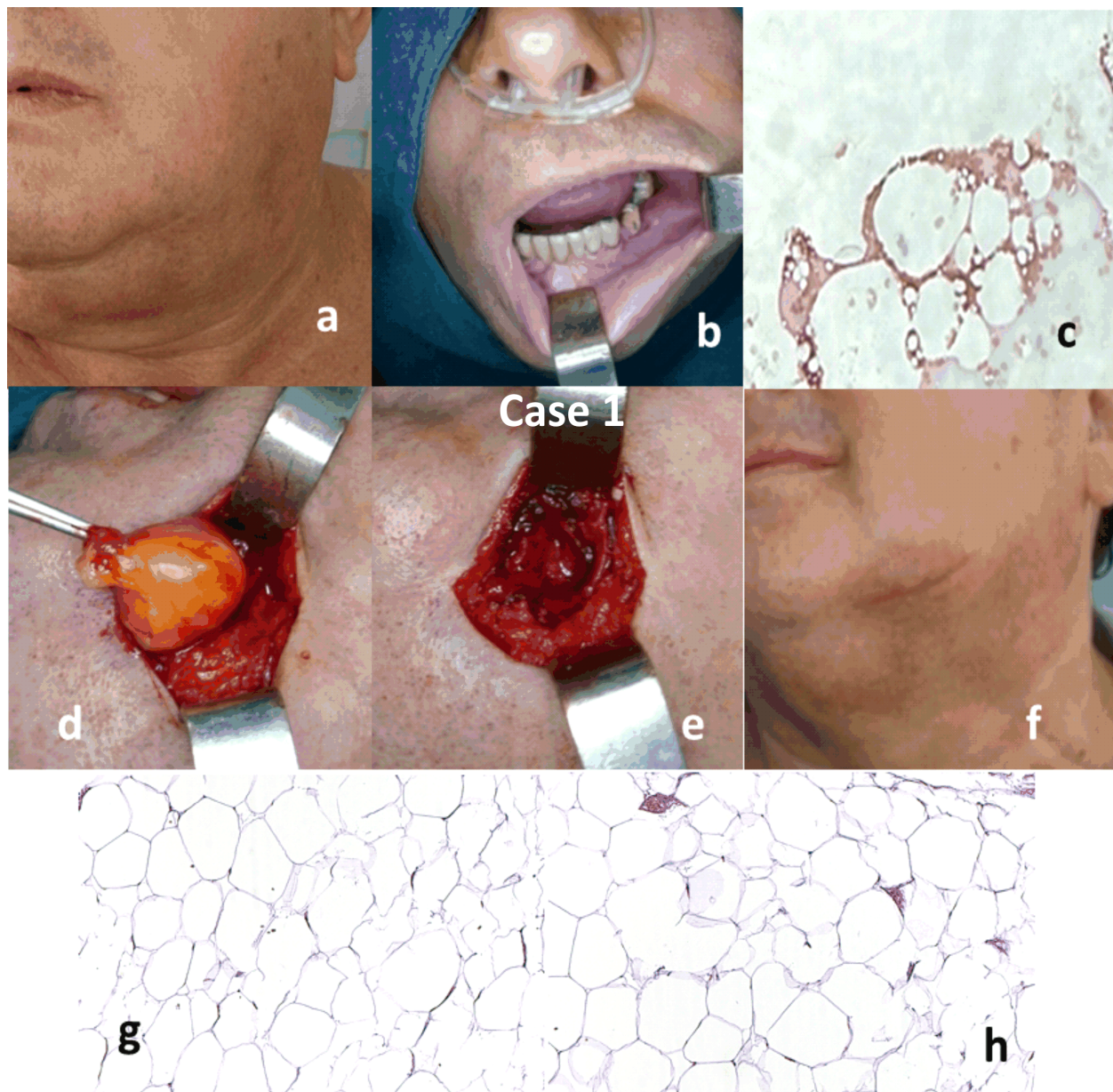


Figure 1 – Case No. 1: Initial aspect showing swelling of the left anterior cervical perimandibular region (a); Intraoral view showing the lesion in the lower left vestibule (b); Papanicolaou-stained smears revealing a monotonous population of large chromophobe cells with eccentric nuclei (c); Intraoperative view (d and e) and postoperative view (f); Adipose tissue with fine connective septa (g and h). HE staining: (c, g and h) $\times 200$.

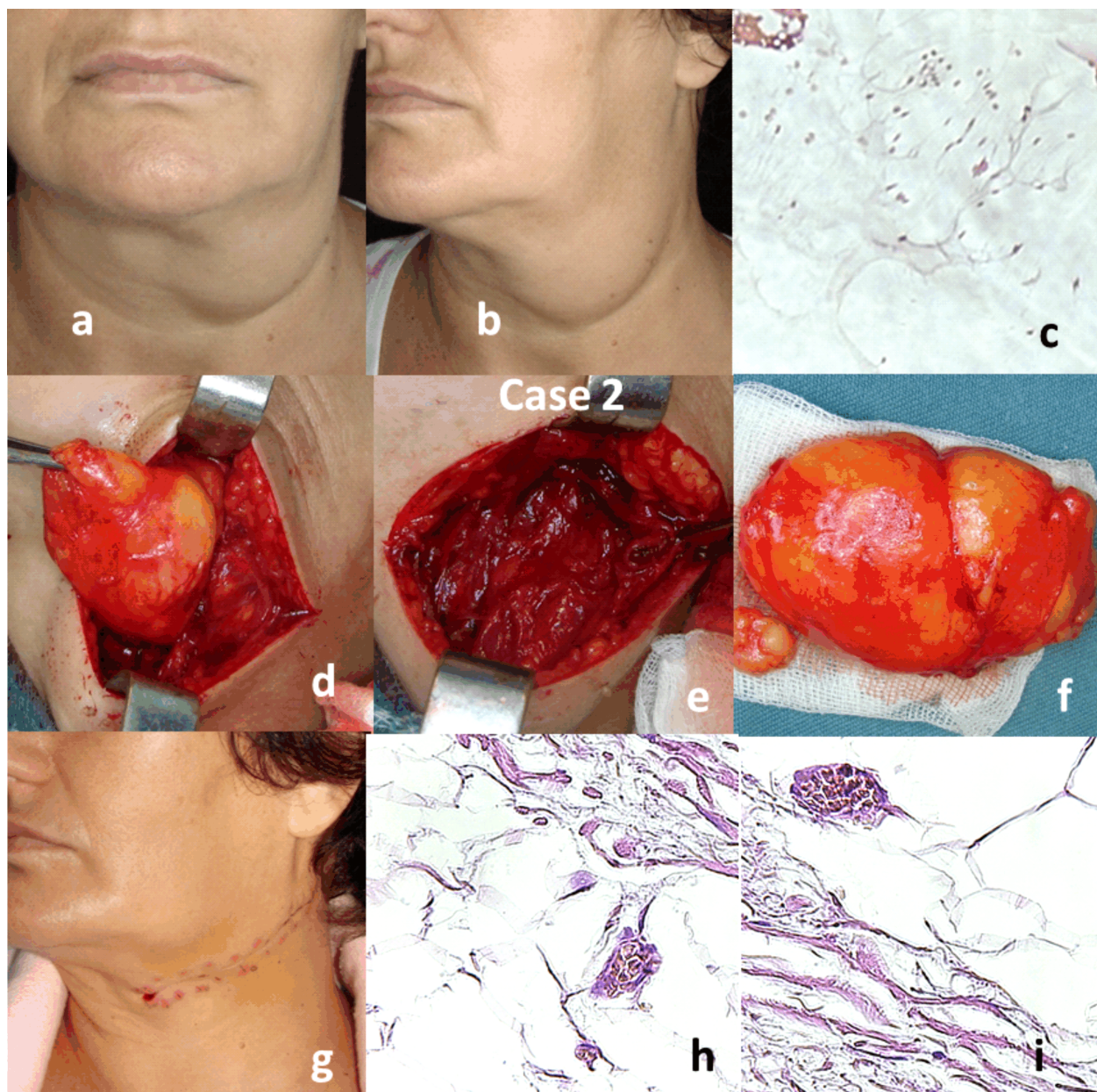


Figure 2 – Case No. 2: Initial front and left lateral view (a and b); Tumor aspirated material revealed many fibroblasts with thin cytoplasmic prolongations (c); Intraoperative view of the tumor (d and e); The macroscopic aspects of the excised tumor (f); Postoperative view of the patient (g); Lobules of mature adipocytes with fibrous connective septa in between (h and i). HE staining: (c, h and i) $\times 400$.

Case No. 3

Clinical data

A 44-year-old man presented with slow-growing painless masses, one in the right and left submandibular and submental region and another one in the retrocervical region. The lesions appeared five years before and grew slowly. No symptoms were noted until recently, when mild discomfort appeared that, including aesthetic reason addressing the patient to the physicians. He was a smoker of 15 cigarettes/day, and a moderate drinker of alcohol. The patient described no other significant personal and collateral past medical history.

On clinical examination, there was a well-defined, smooth, mobile, soft tissue mass, located bilaterally submandibular and submental, 18/6/5 cm in size, and

another one separated retrocervical mass, 6/8/7 cm in size, with normal surrounding skin (Figure 3, a and b).

Intraoral examination showed an intact mucosa, without changes in floor of the mouth or tongue.

Imagistic investigations

CT scan revealed an oval-shaped smooth mass of fat density, measuring 18/6/5 cm, located bilaterally submandibular and submental, without connection to the mandible, and a retrocervical tumor, about 6/8/7 cm. No mediastinal involvement was observed.

Laboratory tests

Triglycerides level was high (308 mg/dL, compared to the normal range between 50–200 mg/dL). In addition, a high glucose level of 174 mg/dL (normal range, 74–106 mg/dL) was registered. TSH level was normal.

Cytological examination

The cytological examination of both tumors of Case No. 3 showed similar aspects consisting in an admixture of mature adipocytes and elongated fibroblasts in a bloody background that supported the diagnosis of fibrolipoma (Figure 3, c and d).

Intraoperative aspects

Surgery consisted in two stages removal of the fat tissue. The first stage of the treatment was performed under general anesthesia (endotracheal intubation) and involved the removal of the lesion from the right and left submandibular regions and from the submental region. After three months, the second stage of treatment was performed under intravenous sedation completed with local anesthesia. At that time, the surgery consisted in the removal of the tumor from the retrocervical region. In

both stages, the tumors were readily dissected manually. For the retrocervical region, the patient was positioned in lateral decubitus and dissection was difficult due to limited access (Figure 3e).

Intraoperative gross examination of excised specimen showed a yellowish mass, with a lobulated surface (Figure 3f). The lesions were sent for microscopic examination.

Histopathological examination

Both tumors were composed of lobules of mature adipose tissue, separated by connective septa that contained large blood vessels (Figure 3, g and h) that highlighted the diagnosis of fibrolipoma.

Taking in consideration the clinical data, the imagistic investigations, the laboratory tests results and the patient's behavior, mainly referring to the alcohol consumption, the lesions were classified as type I Madelung's disease.

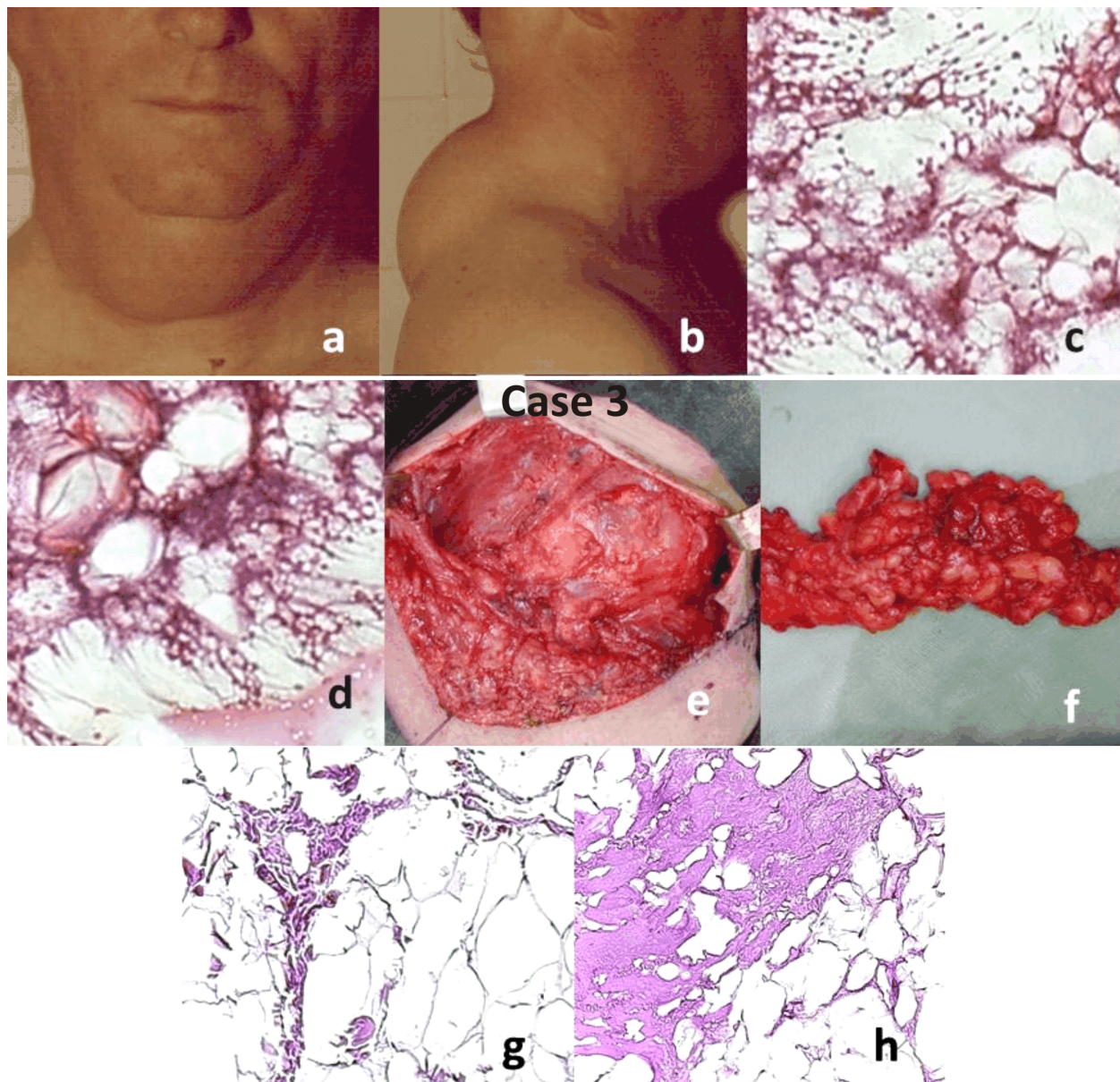


Figure 3 – Case No. 3: Initial front view (a) and right lateral view (b) showing large tumors in submandibular, submental and retrocervical areas; Papanicolaou-stained smears revealing a cellular population composed of adipocytes and fibroblasts in a bloody background (c and d); Intraoperative view of the tumor (e); Non-encapsulated tumor composed of multiple lobules with different sizes (f); The tumors were composed by adipose tissues, thick connective septa with hyalinized collagen bundles (g and h). HE staining: (c, d, g and h) $\times 400$.

Case No. 4

Clinical data

A 45-year-old man presented with multiple lumps in the cervical area. The largest swelling was in the submandibular and submental anterior cervical area and measured 19/7/6 cm. In the retrocervical region, two swelling were noted, one in the left occipital area, 6/7/5 cm in size, and another one in the right occipital area, 6/5/5 cm in size. The increase in neck volume was observed for the last six years. In the last year, the patient reported the presence of mild dyspnea and dysphagia.

He was a hard smoker (30 cigarettes/day), and a heavy alcohol drinker.

The physical examination showed an enlargement of the submandibular and submental anterior cervical area and two occipital masses, 6/7/5 cm in size in the left side of the neck and 6/5/5 cm in size in the right side (Figure 4, a–d).

Imagistic investigations

A facio-cervical and thoracic CT scan showed a fatty accumulation in the referred areas (Figure 4e) and no mediastinal involvement.

Laboratory tests

Triglycerides level was high (320 mg/dL, respectively, compared to the normal range between 50–200 mg/dL). TSH level was normal.

Cytological examination

The cytological examination of the aspired cells from the submandibular and submental lesion revealed that the tumor was composed by predominately by mature adipocytes with clear cytoplasm and eccentric located nuclei mixed (Figure 4f). In contrast, the both occipital tumors revealed many mature adipocytes in a bloody background (Figure 4, g and h). Very few fibroblasts and fibrocytes were observed on the smears from anterior cervical tumor, while small number of them was noticed on occipital tumors smears.

After cytological examination, the diagnosis of presumption for anterior cervical tumor was lipoma and for both occipital tumors, angiolipoma.

Intraoperative aspects

The deep and superficial fatty masses of the neck were excised in three consecutive stages: right anterior cervical, left anterior cervical and retrocervical.

For the right and left anterior cervical masses, surgery was conducted under general anesthesia (endotracheal intubation) and consisted in large horizontal skin incisions, 4 cm below the lower border of the mandible, dissection of the subcutaneous tissues, and exposal and radical removal of the fatty mass. Local hemostasis and wound closure in layers were performed.

For the retrocervical mass, intravenous sedation completed with local anesthesia was used. After the skin incision, the fatty mass was readily dissected manually. Wound closure was performed in layers (Figure 4, i–m). The lesions were sent for microscopic examination.

Histopathological examination

The anterior cervical tumor was composed of lobules of rounded adipocytes with ample, clear, chromophobe cytoplasm and eccentric located nuclei. On the slides from the anterior submandibular and submental tumor of the Case No. 4, the islands of adipocytes were separated by fine collagen fibers admixed with fibroblasts and fibrocytes and containing small capillaries, features consisting with the diagnosis of lipoma (Figure 4n).

In the both occipital region tumors of the Case No. 4, in connective septa between the lobules of adipocytes there were noticed medium and small sized blood vessels with mild or moderate hyperemia, microscopic characteristics of angiolipoma (Figure 4, o and p).

Taking in consideration the clinical data, the imagistic investigations, the laboratory tests results and the patient's behavior, mainly referring to the alcohol consumption, the lesions were classified as type I Madelung's disease.

Post-operative care

After surgery, surgical drains were placed on all cases. Antibiotic, anti-inflammatory and antalgic medication was administrated. None of the patients experienced any post-operative complications such as hematoma or seroma. After discharging from the hospital, the patients came regularly for ambulatory follow-ups.

For the patients with a history of alcohol abuse, our recommendation was to abstain from alcohol consumption.

Follow-up

After surgery, the recovery was optimal for all the patients without any complications. No recurrences were noted during two years of follow-up.

All the data referring to the cases were concluded in Table 1.

Discussions

Lipoma is a benign subcutaneous and submucosal tumor. Head and neck region is the site where a quarter of body's lipomas develop. Despite the data of the literature that describe the most lipomas of the neck region as being located posterior and laterally, and very few cases in the anterior neck, oral cavity, larynx, pharynx, tonsillar or parotid area, in all our cases the lipomas were predominantly developed in the anterior neck. Cheeks and tongue are the predominant localizations for half of the cases of head and neck lipomas, but they may also appear in any soft areas [2].

Symmetric accumulation of fat deposits around the neck is a feature consistent with Madelung's disease. The disease is very rare, the exact incidence being unknown, with a prevalence of one out of 25 000 males in Italy; around 400 cases were published until now in the literature, none of them in the Romania. Orpha.net, European database for rare diseases, cites the syndrome as a rare disease [7, 8].

As in our cases, the disease affects predominantly men, with 20:1 male female ratio and more frequently in the Mediterranean countries.

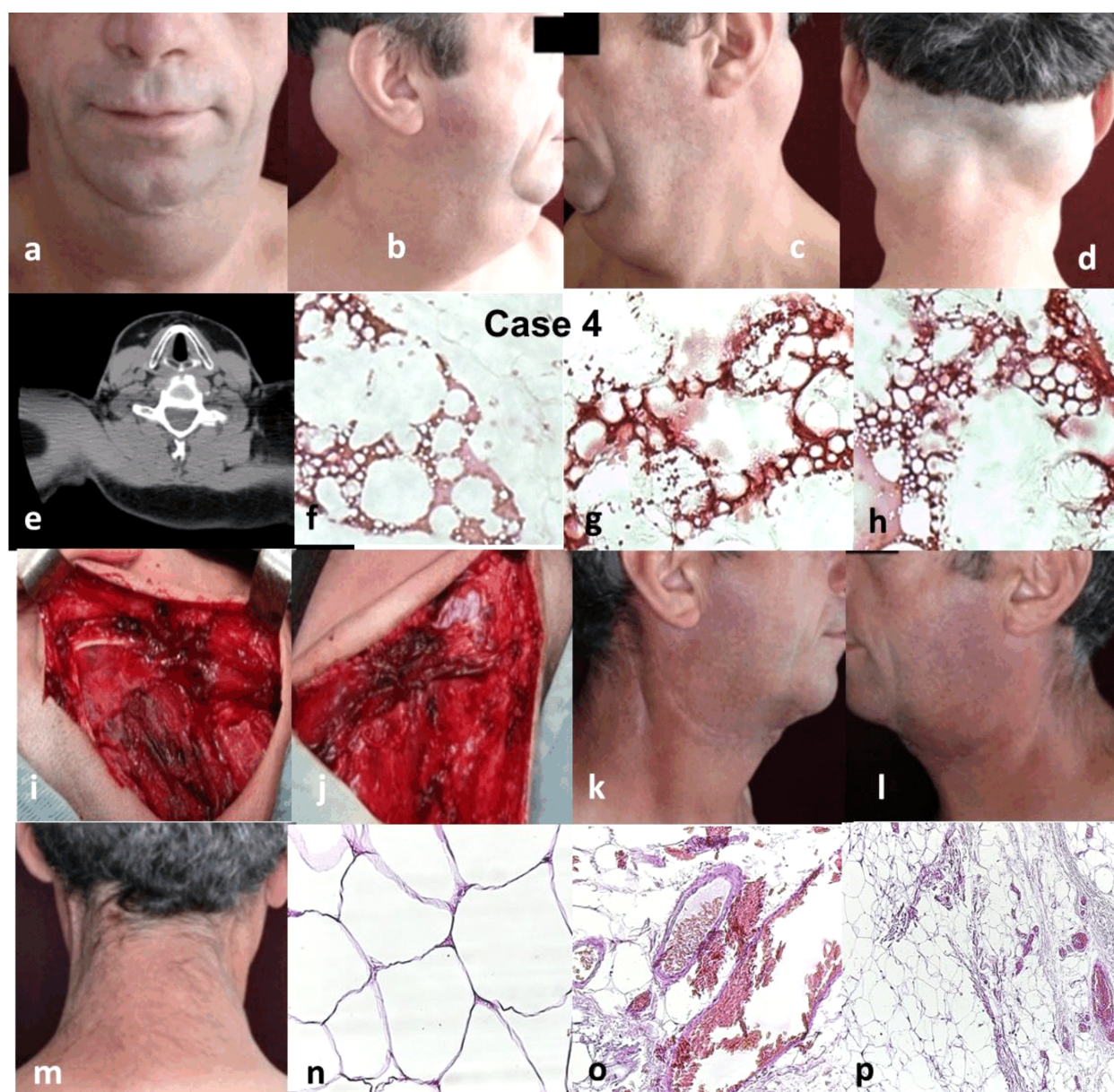


Figure 4 – Case No. 4: Initial view showing an enlargement of the submandibular, submental and occipital areas (a–d) without mediastinal involvement (e); Papanicolaou smears of aspirated material from anterior cervical region tumor showed predominantly mature adipocytes and few erythrocytes (n), while the background of both occipital region tumors showed many erythrocytes (g and h); Intraoperative views (i and j) and postoperative right lateral, left lateral and back views (k–m); HE stained slides of anterior cervical tumor showed mature adipocytes with fine connective septa containing small capillaries (n), while slides of both occipital tumors highlighted many medium-sized vessels in the connective septa (o and p). HE staining: (f–h; o and p) $\times 100$; (n) $\times 400$.

Table 1 – Clinical, radiological and histopathological data of the patients

Parameters	Case No. 1	Case No. 2	Case No. 3		Case No. 4		
Age [years]	57	55	44		45		
Gender	Male	Female	Male		Male		
No. of tumors	One	One	Multiple		Multiple		
Location of the tumors	Left anterior cervical perimandibular	Left anterior cervical perimandibular	Right and left submandibular and submental	Retrocervical	Right and left submandibular and submental	Left occipital	Right occipital
Size [cm]	4/3/3	5/5/4	18/6/5	6/8/7	19/7/6	6/7/5	6/5/5
HP diagnosis	Lipoma	Fibrolipoma	Fibrolipoma	Fibrolipoma	Lipoma	Angiolipoma	Angiolipoma
Time of the evolution [years]	4	5	5		6		
Symptoms	Painless, soft consistency	Painless, soft consistency	Well-defined, smooth, mobile, with normal surrounding skin, painless		Painless		
Reasons for medical addressability	Aesthetic	Aesthetic	Aesthetic, low mobility of the neck		Aesthetic, low mobility of the neck, dyspnea		

Parameters	Case No. 1	Case No. 2	Case No. 3	Case No. 4
<i>Clinical diagnosis</i>	Lipoma	Lipoma	Madelung's disease	Madelung's disease
<i>Imagistic investigation</i>	Panoramic radiograph	CT scan	CT scan	CT scan
<i>Medical history</i>	–	–	Diabetes mellitus; obesity; hypertension	Hypertension
<i>Laboratory tests</i>	Without significant changes	Without significant changes	Triglycerides level of 308 mg/dL [*] ; Glucose level of 174 mg/dL ^{**}	Triglycerides level of 320 mg/dL
<i>Observations</i>	–	–	Alcohol abuse; 15 cigarettes/day	Alcohol abuse; 30 cigarettes/day

HP: Histopathological; CT: Computed tomography; *Triglycerides normal range: 50–200 mg/dL; **Glucose normal range: 74–106 mg/dL.

Usually, the adipose benign tumors cause few problems other than those of localized mass. As in our cases, most tumors tend to grow insidiously, without symptoms. In all our cases, the tumors were painless and soft. Despite published data, in our cases, there were no pathological signs on the overlying skin (changes in color or vascularization), nor cervical lymph nodes enlargement, as cited in the literature [2].

In most cases, isolated lipomas are well circumscribed, encapsulated and lobulated, while the fat deposits of Madelung's disease have ill-defined borders.

Even if the patients are diagnosed with isolated adipose tumor or Madelung's disease, the main reason for medical addressability, as in our cases, is the aesthetic problem.

After years of development, fat stores reach enormous sizes and, in advanced cases, dyspnea, dysphagia and dysphonia occur [1]. Occasionally, the lipoma may invade muscles or grow between muscles, cervical fascia, and vascular-nervous elements (the infiltrating lipoma) [9, 10]. Although the disease is considered benign, involvement of the oral cavity (tongue) and mediastinum sometimes appear, leading to tracheal or superior vena cava compression syndromes [1], situations not detected in our study.

In 80–99% of cases, there were noted arthralgia, joint stiffness and gait disturbance.

In our cases, other symptoms besides esthetic problems included only reduction of the neck mobility in two cases and dyspnea in one case.

The disease is associated with other co-morbidities as hepatomegaly (30–79%), diabetes mellitus (30–79%), hyperuricemia, peripheral polyneuropathy (30–79%), hypothyroidism and malignant tumors of the upper airways [9, 10]. In our cases, the co-morbidities diagnosed were diabetes mellitus (one case – 25%), obesity (one case – 25%), hypertension (two cases – 50%), triglycerides elevation (two cases – 50%), chronic alcoholism (two cases – 50%).

In the pathogenesis of Madelung's disease was cited an enzymatic defect of lipid metabolism and a mutation of mitochondrial deoxyribonucleic acid (DNA) involving mitochondrially encoded transfer ribonucleic acid (tRNA) lysine (*MT-TK*) gene [9]. It seems that alcohol consumption can be a trigger for the disease, 80% of the patients having associated alcoholism [10, 11]. Rarely, more than one family member is affected that suggest the possibility of disease inheritance, even if the exact mechanism was not fully demonstrated. In our cases, none of the patients presented family medical history of the disease.

The diagnosis of Madelung's disease is based on physical examination and imaging studies. CT and magnetic

resonance imaging (MRI) are very useful to define the positive and differential diagnosis of cervicofacial lipomas. Composed of mature adipose tissue, classic lipomas appear as homogeneous hypoattenuated masses, with similar radiological aspects to those of subcutaneous fat. The lesions shape is more variable with depth because of the surrounding structures that can compress and deform them [12, 13].

Because of its better soft tissue contrast resolution, MRI represents the gold standard for tumor delineation, location and extent. MRI allows highlighting cleavage plans between the lipoma, muscle, and vessels [14]. These aspects are of high importance in the oral-facial region because in this region, the lipomas are delineated by a very thin capsule being surrounded by normal fat tissue. Using of intravenous contrast agents is important when sarcomatous degeneration is suspected to better highlight the tumor margins and irregular vascularization [13, 14].

Only in few cases, a biopsy is needed in order to complete the diagnosis of Madelung's disease. Lipoma, angiolipoma and fibrolipoma are benign variants of lipomatous lesions that have been described. Lipoma is well vascularized, but the vascular network often is compressed by the distended adipocytes. Angiolipoma is characterized by prominent vascular components. Fibrolipoma is diagnosed when between the adipose cells can be observed high quantity of fibrous tissue comprising of collagen fibers, fibrocytes and fibroblasts. The fibrocytes and fibroblasts must be normal, without atypia.

All the benign variants of lipoma have been diagnosed at the patients from the present study. All the tumors were composed of mature adipocytes. Even if in the literature is cited that adipocytes form Madelung's disease, tumor exhibit an increased, tumor-like, proliferation rate, in our cases no mitoses were identified [15]. In addition, some authors showed that the adipocytes of Madelung's disease lipomas have intermediate characteristics between mature adipocytes and lipoblasts observed in liposarcomas, none of these was observed in our cases [15].

On histopathological grounds, the most lipomas are easy to diagnose. Even so, there are some types that can cause diagnostic problems, including infiltrating, pleomorphic and spindle cell lipoma, and lipoblastoma [1, 15–17].

The differential diagnosis of isolated lipoma of the head and neck area include neurofibromatosis, schwannoma, different forms of muscular dystrophy and lymphoproliferative disorders. Madelung's disease lipoma could be also misdiagnosed as thyroid disease, neurofibromatosis or sarcomatous tumors. When the oral cavity is involved, achromic melanoma and schwannoma of the oral mucosa are between differentials [18–29].

The management of Madelung's disease includes the surgical removal of the lesions or ultrasound-assisted liposuction. Lipomas are typically easy to remove, but in many cases, it is not possible to remove them entirely. Therefore, regardless the treatment method, surgical removal or liposuction, the tumors can recur [30–32]. The recurrence incidence varies among studies but, the high rate of recurrence (60% of the head and neck cases) reported by some authors is perhaps attributable to the difficulties in radical excision [16].

At the cases diagnosed with Madelung's disease, the correction of associated metabolic disorders, weight loss, cessation of the alcohol intake and smoking should complete the therapeutic scheme [9]. Even so, these supplementary treatment procedures have not statistically reduced the rate of recurring.

Recently, intratumoral mesotherapy with phosphatidylcholine of Madelung's tumors was reported as potential treatment [9, 30–32].

Rarely, the lipomas can acquire malignant transformation, only two cases of liposarcoma being presented in the English literature, one in 59-year-old Italian woman with synchronous breast cancer and another one in a 57-year-old woman after seven years of disease evolution [9, 10]. Any changes in the clinical appearance as asymmetry, change of color or consistency, signs and symptoms of compression, should address the patient to the physicians for a prompt surgical removal of the fat deposits or at least performing of a biopsy.

☒ Conclusions

The present study emphasized the clinical, histopathological and therapeutic options of Madelung's disease, a rare disorder, relatively underdiagnosed. It also highlighted that is no difference between Madelung's disease and common lipoma from the cytological and histopathological point of view, the correct diagnosis of the former lesion being established only in the correlation with clinical, imagistic and laboratory data. Moreover, the correct diagnosis of Madelung's disease is essential in order to add other treatment options to the surgery or liposuction, as avoiding alcohol or medication for metabolic disorders.

Conflict of interests

The authors declare that they have no conflict of interests.

Author contribution

Marius Octavian Pricop and Nicolae Constantin Balica equally contributed to the article.

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Received: November 27, 2013

Accepted: August 29, 2018