CASE REPORT

Skull base tumor invading both cavernous sinuses. Adenoid cystic carcinoma mimicking a meningioma

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Abstract

We present a case of an anterior skull base tumor invading both cavernous sinuses and extending into the right orbit in a 55 years old female. The radiological aspect was confusing, being highly suggestive for an extensively invasive meningioma. However, the orbital portion of the tumor, which was surgically removed, proved to be an adenoid cystic carcinoma. Some peculiar immunohistochemical findings were obvious, as well as a lack of continuity of the tumor with the local lacrimal gland. This is an unusual situation, when a lacrimal gland tumor spread along the cavernous sinus, cross the midline and approaches the contralateral orbit. Such local extension should be considered in the differential diagnosis of anterior skull base tumors.

***Keywords*:** meningioma, lacrimal gland, adenoid cystic carcinoma, bilateral invasion, cavernous sinus.

Introduction

Skull base tumors represent a surgical problem by their histological variety and complex location. In such tumors, cases of cavernous sinus syndromes can occur and engender difficulties in diagnosis. The majority are generated by intracranial meningiomas, which have a tendency to local extension along the optic nerve sheath. Conversely, orbital meningiomas could spread intracranially, on the same way [1].

Other much rare tumor with tendency to invade the cavernous sinuses is the adenoid cystic carcinoma (ACC). The imaging examinations can be confusing between the two entities [2–4].

We present an ACC case with an exceedingly large intracranial extension and bilateral cavernous involvement. Such an unusual case is not reported to date, to our knowledge.

Patient and methods

A 55-years-old woman was admitted in our hospital with visual impairment, predominantly on the right side, and diplopia, progressively installed starting three years ago. The computed tomograph examination revealed a 2/3 cm tumor of the left cavenous sinus.

The tumor was surgically removed and diagnosed as meningothelial meningioma, based only on routine histopathology. An improvement in her condition was observed for approximately three months, the diplopia recurring afterwards.

At this stage, a neurological examination revealed oculomotor and trochlear nerves paresis, trigeminal paresthesia, a marked loss of vision in eyes, bilateral exophtalmia and a left pyramidal syndrome. This time, the CT-scan image suggested a meningioma "en-plaque" located on the external aspect of the right cavernous sinus, sphenoid jugulum, and right orbit, also extending to the lateral aspect of the left cavernous sinus (Figure 1, a and b).

MRI examination confirmed these findings. The global image was that of a suprasellar tumor largely invading the skull base on both sides on the internal aspect of the bone.

The patient underwent a new surgical procedure, which also visualized the extention into the right orbit. Intraoperatively, the tumor was a gray mass, richly vascularized, extending from the left to the right cavernous sinus.

Due to the extreme extension of the tumor mass, an orbital exenteration was not performed and only a portion of the tumor was removed, mostly for diagnostic and decompressive purposes. Postoperatively, the patient was discharged with a stable condition, with an indication for oncological survey and complementary therapy. At routine microscopically examination the first surgical sample, as well as large areas of the second one, showed aspects highly suggestive for a meningioma.

The tumor cells were grouped in nests, with a meningothelial appearance and lacking observable membranes, surrounded by a fibrous, collagenous stroma (Figure 2).

At the second intervention these aspects were also present but, on other areas, typical features of an ACC

were obvious, i.e. small cells with angular profiles with minute cysts within cell groups (Figure 3).

Immunohistochemical analysis of the case was performed, using antibodies against: cytokeratins KL1 (Immunotech, 1:200 dilution), CK7 (Dako, Glostrup, 1:50 dilution), CK14 (Novocastra, 1:50 dilution) and 34betaE12 (Dako, Glostrup, 1:50 dilution), S100 protein (Dako, Glostrup, 1:500 dilution), smooth muscle actin (Sigma, 1:4000 dilution), CEA (Dako, Glostrup, 1:50 dilution), EMA (Dako, Glostrup, 1:75 dilution), vimentin (Biogenex, 1:100 dilution), E-cadherin (Dako, Glostrup, 1:50 dilution), CD44 (Novocastra, 1:50 dilution), CD54 (Novocastra, 1:50 dilution), CD117 (Dako, Glostrup, 1:250 dilution) and Ki67 (Dako, Glostrup, 1:50 dilution).

The tumor cells were positive to all epithelial markers (KL1, 34betaE12, CK7, CK14, and EMA), vimentin, smooth muscle actin, and CD44 on restricted areas (Figures 4–6).

A striking aspect was the positivity for CD117 (c-kit) in many cell groups (Figure 7) and for E-cadherin in tubular structures (Figure 8).

CD54 was negative in tumor cells. The Ki67 labeling index was relatively low, about 5%.

Discussions

Anterior skull base tumors can be challenging for the neurosurgeon as well as for the pathologist. Among these, meningioma is the most frequently encountered, with about 20% of cases [5].

This tumor has the propensity to invade the orbit or, conversely, to originate from the optic nerve sheaths and subsequently extend intracranially [6].

Adenoid cystic carcinoma (cylindroma) is a much rarer entity that could also have the latter behavior. ACC has the origin in the salivary, lacrimal, and other exocrine glands, and is rare in neurosurgical practice [7].

Some radiological and histopathological features encountered in our case made difficult its correct identification. A vague histological resemblance with a meningothelial meningioma, mostly at first resection, produced confusion and generated an erroneous diagnosis. Even with the immmunohistochemical assessment, the positivity for EMA and vimentin in most tumor cells could induce the false diagnosis of meningioma, since vimentin is also commonly expressed in ACC [8].

However, the rest of the antibody panel prompted us a correct diagnosis. Intracranial forms of ACC are considered as being rare, even though recently more than 100 cases have been described [10].

In many cases, the correct primary of such intracranial ACC can be difficult to define. Even primitive intracranial cases are described [9] but which have to be regarded with caution.

In our case the lacrimal gland is the most probable origin, even though it was not macroscopically involved.

The positivity for different antibodies is in accord with other localizations of the tumor. Cytokeratins are currently expressed in ACC. Smooth muscle actin is also known to be present in myoepithelial cells originating from the gland [8].

The presence of CD44 in many cells do not seems to be related to local invasion or global behavior, at least in ACC with salivary gland origin [11].

A reduced expression of E-cadherin is described in progressive forms of ACC [12].

In our case, despite the extreme extension of the tumor, E-cadherin was positive on large areas. The reaction to CD117 (c-kit) can be encountered in 25–78% of various ACC reports, even though without prognostic significance [13, 14].

The unusual extension, with bilateral involvement of the cavernous sinuses is difficult to be explained by the intrinsic tumor cells characteristics. The low Ki67 labeling index do not guarantee a benign behavior, because of the unpredictable manner this tumor type has to locally extend, destroy and even metastasize to remote places.

Even though the local extension is large, in some cases a palliative surgery could ensure a long-term survival [15].

Due to the relative uncommonness of this tumor type, the optimal neurosurgical and oncological attitude (radiotherapy and chemotherapy) in particular cases remain to be determined [16].

Conclusions

Adenoid cystic carcinoma of the skull base can generate some difficult problems regarding its precise origin, pathological identification and appropriate therapy. A bilateral extension of such a tumor is first time reported here, to our knowledge. Neurological surgeons and pathologists must be aware of the possibility to be confronted with this particular tumor type.

References

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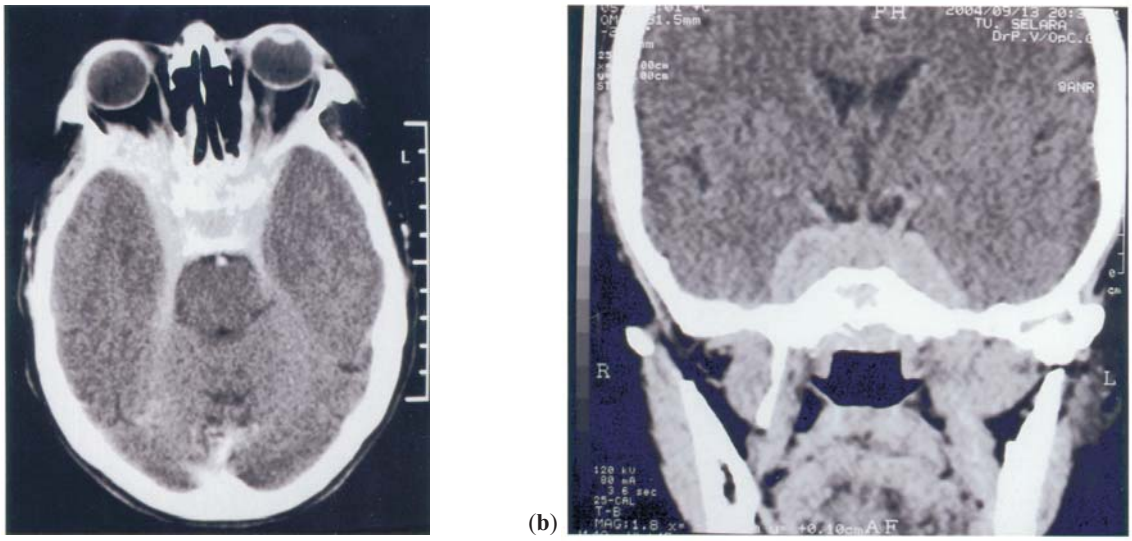

Figure 1 – *Computed tomograph images of the tumor: (a) spontaneous hyperdense mass is present in the median regions of the cranial base, extending into the right orbit; (b) on coronal section the tumor is located in the suprasellar zone*

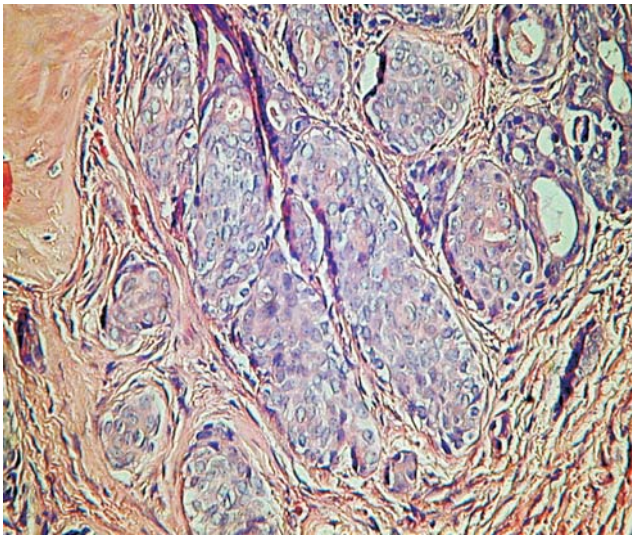

Figure 2 – *The tumor cells are arranged in nests surrounded by a fibrous stroma. The image highly suggest a meningioma (HE staining, ×400)*

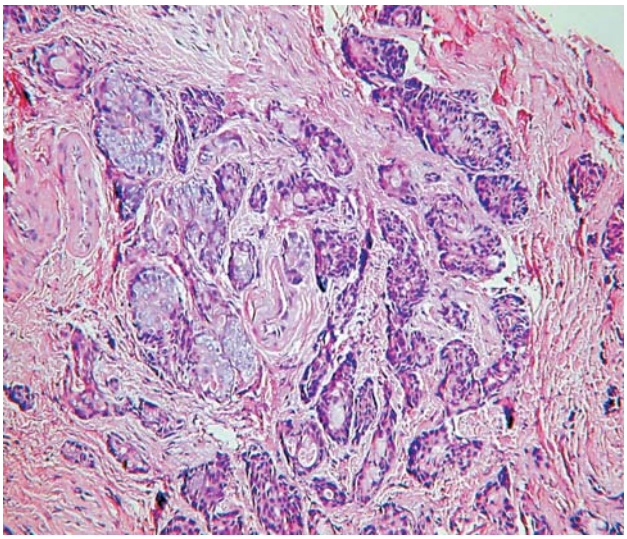

Figure 3 – *Close to some features from the previous image, large areas are conspicuously those of an adenoid cystic carcinoma (cylindroma) (HE staining, ×200)*

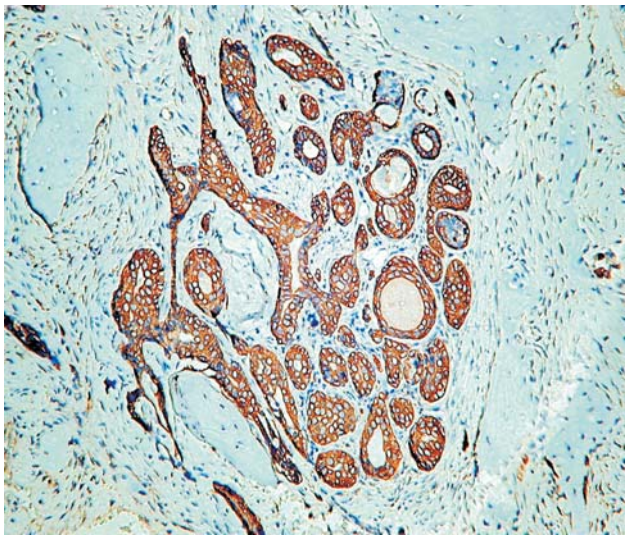

Figure 4 – *CK14 is strongly expressed by the tumor cells (IHC staining, ×400)*

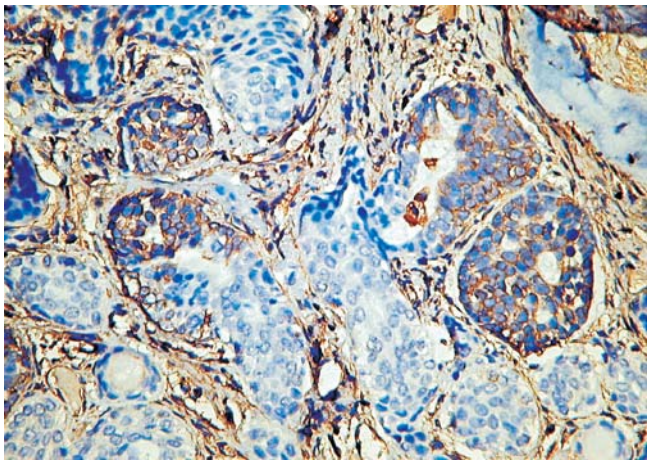

Figure 5 – ***Vimentin is positive in some group of cells, mostly in those resembling meningioma (IHC staining, ×400)***

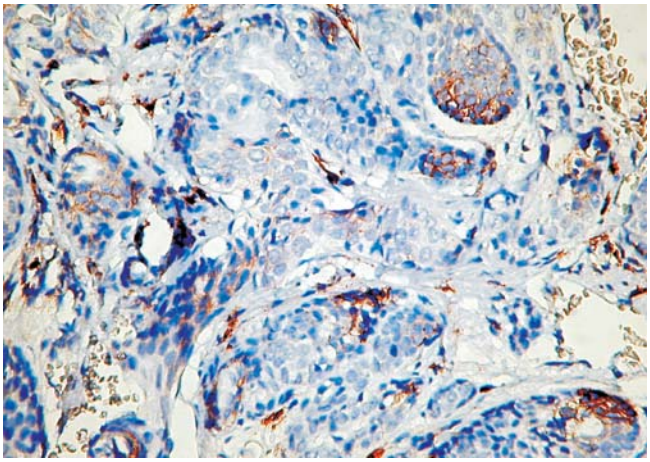

Figure 6 – ***CD44 expression is limited to some small cell clusters (IHC, ×400)***

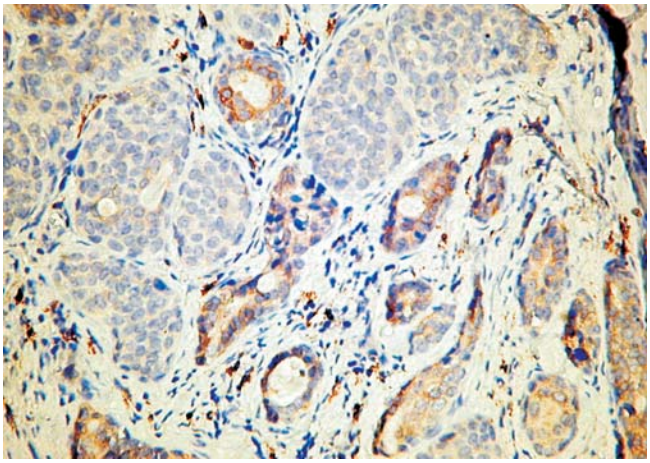

Figure 7 – ***CD117 has various positivities, more intense in the cells typical for adenoid cystic carcinoma and more pale in the "meningothelial" islands (IHC staining, ×400)***

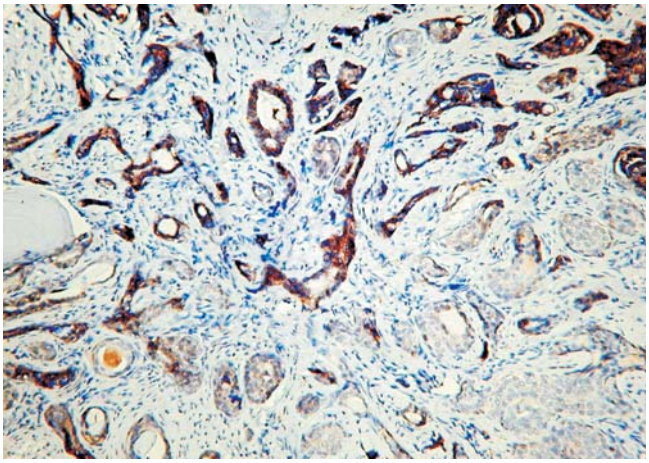

Figure 8 – ***E-cadherin appear positive mostly in tubular structures (IHC staining, ×200)***

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