CASE REPORT

An intrajugular paraganglioma. Unusual presentation of a classical tumor

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Abstract

Paragangliomas arise from the extraadrenal neuroendocrine system. They are locally aggressive tumors, causing adjacent invasion, bone destruction and compression related symptoms. We present a 35-years-old woman with a peculiar paraganglioma lacking all these features, and strictly located within the jugular vein. Differential diagnosis is detailed since other entities could have dissimilar clinical behavior. To the best of our knowledge, this is a very unusual site of occurrence for paragangliomas, and only two other comparable cases have been described.

***Keywords*:** paraganglioma, intrajugular, immunohistochemistry, prognosis, differential diagnosis.

Introduction

Paragangliomas are tumors of the specialized extraadrenal neuroendocrine system. The overall location of paragangliomas is in accordance to the sites of normal paraganglia: the carotid body, the jugulotympanic body, the vagal body, etc. If the jugular vein bulb is concerned, they are called glomus jugulare tumours. The prevalence is low, paraganglioma accounting for only 0.6% of neoplasms of the head and neck region [1]. Their classical evolution is toward local invasion, with destruction of the petrous bone, following the low resistance paths, toward mastoid cell tracts [2], vascular channels [3], or Eustachian tube [4].

A strict intravascular localization within the jugular vein is very rarely reported to date to our best knowledge [5, 6]. In our experience, this is the first case with such a particular presentation.

Clinical report

The patient, a 35-years-old woman, was admitted to our hospital with dysphonia, and left facial asymmetry with a 12-months duration. In the last 3–4 days, appeared new symptoms: headache, gait troubles and nausea, as well as dysarthria, dysphagia and dysphonia.

The physical examination revealed paresis signs of most cranial nerves on the left side (VI, IX, X, XI, XII).

A contrast CT-scan showed a tumor mass in the jugular vein superior bulb, with strong contrast enhancement. The MRI revealed the same aspect, of a tumor of 38/35/45 mm with intermediate signal in T1-weighted and hyper-signal in the T2-weighted sequences, with strong enhancement after intravenous administration of gadolinium (Figure 1).

The tumor was removed in block with the corresponding segment of the vein, with good postoperative evolution.

The patient was discharged with no additional therapeutic recommendations.

Follow-up examinations up to one year later disclosed only persistent facial hemiparesis and dysphonia. The MRI examination revealed no tumor recurrence.

Pathologic report

Macroscopically, the surgical material was a portion of the jugular vein, filled with a fleshy mass, the overall aspect being that of a sausage.

Classic histological stains (Hematoxylin and Eosin, Masson's trichrome) were first performed.

Microscopically, the tumor appeared rigorously restricted to the vein lumen, with no invasion of any of the vessel wall structures or of small lymph node, which was also removed (Figure 2).

The tumor had the typical aspect of a paraganglioma, with cells arranged in nests or clusters separated by a rich vascular stroma (Figure 3). No mitoses or necroses were detectable.

Immunohistochemistry was performed on the paraffin-embedded material using the EnVision+ Dual Link System Peroxidase kit (Dako, Carpinteria, CA, USA), according to the manufacturer's instructions. Primary antibodies against the following antigens were used: chromogranin (1:100) (Novocastra, Newcastle Upon Tyne, UK), synaptophysin (1:50), S100 protein (1:500), Ki67 (1:100), CD34 (1:50), FVIII associated antigen (1:50), CD31 (1:40), GFAP (1:50) (Dako, Glostrup, Denmark).

The Ki67 labeling index was counted on 100 nuclei in the most positive areas.

Chromogranin (Figure 4) and synaptophisin (Figure 5) were positive in the tumor cells. S100 protein showed a weak positivity in the chief cells in addition to a more intense one in the sustentacular cells (Figure 6). Ki67 showed a labeling index below 5% (Figure 7). Vascular markers (CD34, FVIII, and CD31) as well as GFAP proved to be negative.

Discussions

Paragangliomas may arise in various locations and have various local extensions. We describe a very unusual case, where the tumor is located strictly within a vessel, with no invasion of the adjacent structures. Only very rare such cases have been described [5, 6]. Some paragangliomas, called "complex" rather by their gravity than histology [7], were described as having sometimes a large vascular extension [8].

In our case, however, the tumor extended only 2–3 cm downstream from the superior bulb of the jugular vein. The histological features were common, with typical positive immunoreactivity. S100 protein was found to be positive mainly in the sustentacular cells. Even though it was also expressed in some of the chief cells, this is not so peculiar, since a certain degree of this pattern is common [9].

An element of clinical importance is the limitation of the tumor proliferation only to the jugular lumen, the vessel wall or adjacent tissues lacking tumor invasion. This allowed a complete resection in this case. A small adjacent lymph node that was also removed showed no signs of microscopical or immunohistochemical presence of tumor cells. The immunohistochemical profile could not permit any assumptions regarding this unusual site of occurrence.

Regarding the prognosis, a low degree of extension within the vessel lumen and the lack of infiltration into the adjacent tissues suggest a good evolution. The tumor being considered as benign by some authors [10], at least in our case the evolution is expected to be good, since large, complete resection was performed. The low Ki67 labeling index, less than 5%, could be in accord to this. Follow-up data at one year after surgery suggest encouraging conclusions (no recurrence visible on MRI).

On the other hand, some other tumors and tumor-related lesions may arise within vessels and generate diagnostic difficulties for the pathologist. Papillary endothelial hyperplasia (PEH, Masson's tumor) is an intravascular endothelial proliferation that sometimes mimics angiosarcoma. This benign lesion could possibly arise in veins with subsequent nervous system symptoms [11].

It should be kept in mind because of its recurrence potential if incompletely resected [12]. Nevertheless, the histological aspect of PEH is completely different, with a papillary growth pattern originating from the vessel walls, it does not express neuroendocrine markers as chromogranin and S100 protein, as did our case, and the vascular markers (CD34, CD31, FVIII) are positive.

Angiosarcoma is another differential diagnosis of a tumor arising within a vessel [13]. In this case, the proliferation is much more extensive and invasive, and has the tendency to be multicentric. A conspicuous mitotic activity is common and neuroendocrine markers are negative. The histological aspects are different and vascular markers are positive.

The intravascular localization of a lesion can also be missdiagnosed as vascular thrombosis [14]. The clinical context (no trauma or pro-coagulative condition, etc.), MRI aspects and most of all the histology ruled out this possibility.

Conclusions

A tumor located within the jugular vein is an unexpected finding for the neurological surgeon.

Paraganglioma is particularly unusual in this location. Its immunohistochemical characteristics must be carefully examined, since its behavior is highly variable. Its presence within the head and neck veins should be considered as a possibility.

Several other tumors or pseudo-tumors must also be kept in mind for this location. In our case, several elements plead for a benign behavior and no additional therapy was applied, with good results, at least during the short follow-up period.

References

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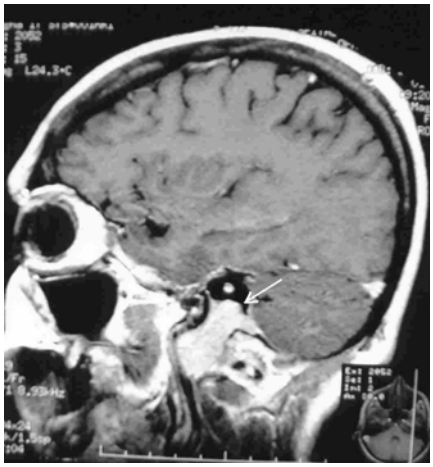

Figure 1 – ***MRI, T1-wheighted aspect of the tumor after gadolinium administration. The mass is located within the jugular vein and has a strong contrast enhancement***

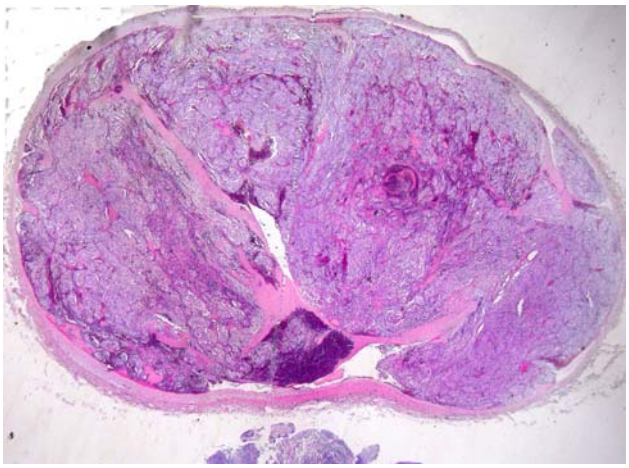

Figure 2 – ***General view of the tumor on microscopic examination. The proliferation is strictly limited to the vein lumen. The small lymph node above is tumor free (HE, ×10)***

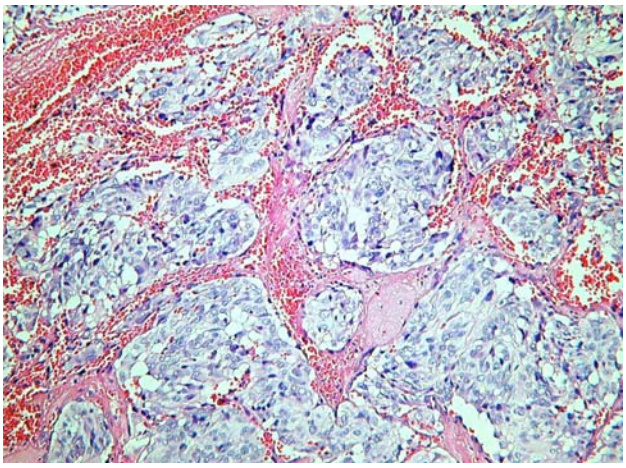

Figure 3 – ***Histological aspect of the tumor. The cells have clear cytoplasm and are arranged in lobules, surrounded by delicate vascular stroma (HE, ×400)***

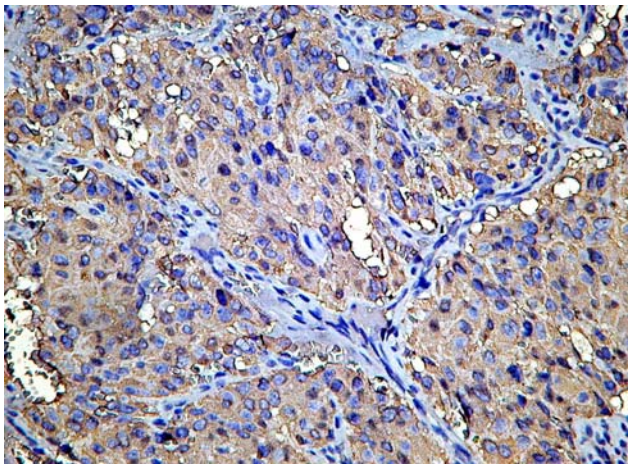

Figure 4 – ***Diffuse tumor reactivity for chromogranin (DAB–peroxidase method, ×400)***

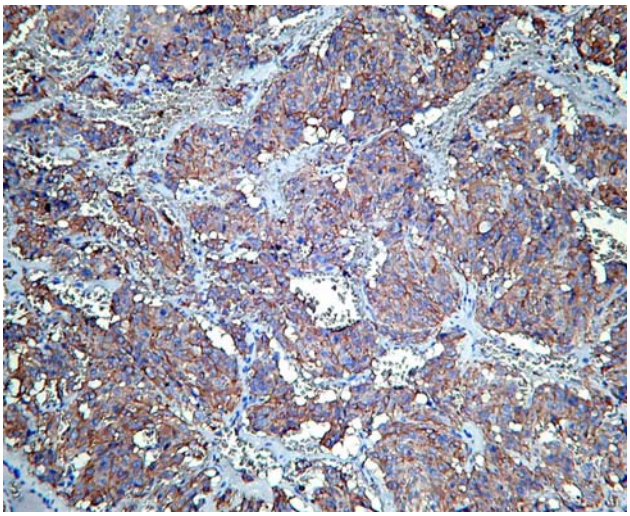

Figure 5 – ***The tumor cells are strongly reactive for synaptophysin. Tumor septa in this region are thicker and clearly devoid of reactivity (DAB–peroxidase method, ×400)***

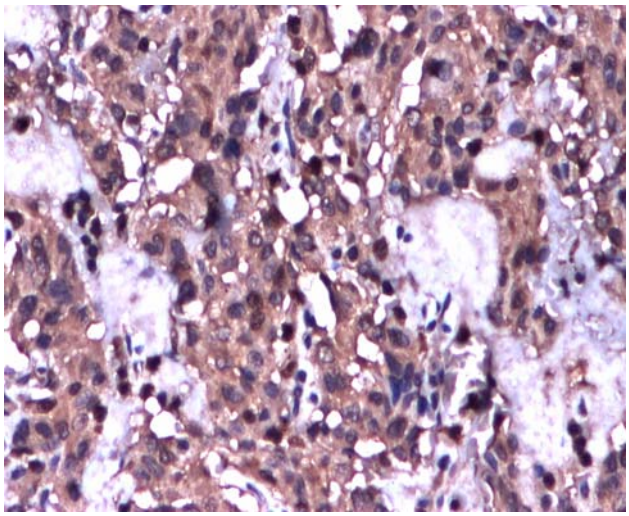

Figure 6 – ***S100 protein is diffusely expressed in the tumor cells, more intensely in the sustentacular ones (DAB–peroxidase method, ×400)***

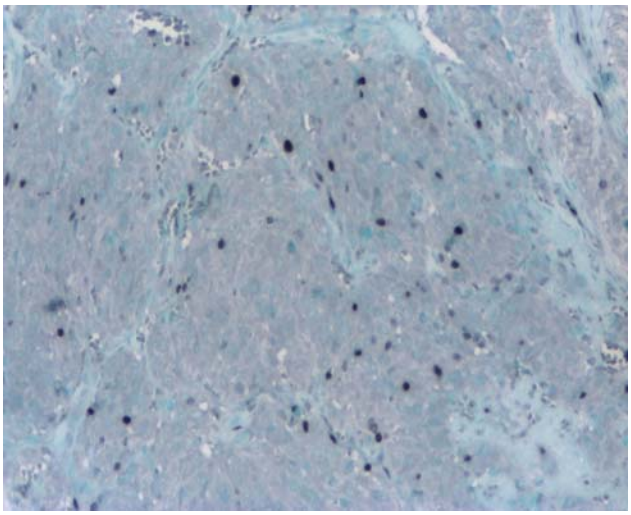

Figure 7 – ***Ki-67 immunolabeling is scarce (DAB–peroxidase method, ×200)***

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