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A case report and review of the literature.

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Intracranial soft-tissue glomus tumor (glomangioma) in a young-woman. A case report and review of the literature.

BACKGROUND: *Glomus tumors, or glomangiomas, are benign vascular tumors typically seen at distal extremities. These tumors differ from paragangliomas and classically present in the female population between the 4th and 5th decade. Intracranial localizations have not been described in literature in the adult population.*
CASE DESCRIPTION: *We present a case of a 32 year-old woman with a 3 months history of progressive left-sided visual loss and headache. A pre-operative MRI showed a homogeneously enhancing lesion extending from the left cavernous sinus to middle cranial fossa at first suspected to be a cavernous sinus meningioma. Eventually, histopathological analysis concluded for a glomangioma diagnosis. Post-operative RT was also performed.*
CONCLUSIONS: *From our experience it is very important for clinical management considering glomangiomas in differential diagnosis of a homogeneously enhancing extra axial mass. Subtotal resection followed by radiation therapy determined no recurrence of the disease up to 7 years.*

KEY WORDS: Glomangioma, Glomus tumor, Soft-tissue tumor

Introduction

Soft-tissue glomus tumors, or glomangiomas, are benign vascular tumors typically seen at the distal extremities, often subungual. These tumors should not be confused with paragangliomas (sometimes referred to as glomus tumor), that arise from non-chromaffin paraganglion cells. Glomus tumors are thought to represent hamartomatous proliferations of modified smooth muscle cells originating from pre-existing normal glomus cell populations. There are two main components on microscopy: branching vascular channels and aggregates of specialised glomus cells [1,6].

Glomangiomas classically present in young to middle aged (4th to 5th decades), with female predilection. They can be multiple in ~10% of cases. Glomus tumors account for 1-5% of the soft- tissue tumors in the hand [2]. A successful resection of these lesions depends on the high vascularization and intraoperative haemorrhagic risk. As such, neurosurgeons, neuropathologists, and head and neck surgeons may be unaccustomed to treating patients with these lesions.

Case Report

A 32 year-old woman presented to our department with a 3 month history of progressive left eye symptoms including diminished intraocular and extra ocular movements, proptosis, visual loss, and headache.
Magnetic Resonance Imaging demonstrated a homogeneously enhancing lesion extending from the left cavernous to middle cranial fossa (Fig. 1). A pre-operative

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angiography was undertaken (Fig. 2). Neuroradiological studies demonstrated a massive lesion with an important vascular component and massive homogeneous contrast enhancing.
At first, it was suspected to be a cavernous sinus meningioma, extending to left middle cranial fossa with implication of left optic canal.
Patient underwent surgical treatment to remove the lesion. It was approached using a left fronto-temporo-parietal craniotomy.
Tumor appeared hyper-vascularized and so resection was performed with removal of small tissue fragments and progressive haemostasis. Only subtotal tumor resection was possible because of the close implication of the ipsilateral cavernous sinus.
Base of the lesion, lying on the middle cranial fossa floor, showed a blood supply from both middle meningeal artery and small arteries originated from skull base bone (Fig. 3).
In postoperative course, it was necessary to place a spinal cerebrospinal fluid drainage because of a skin flap tumefaction, which resolved completely in 7 days.

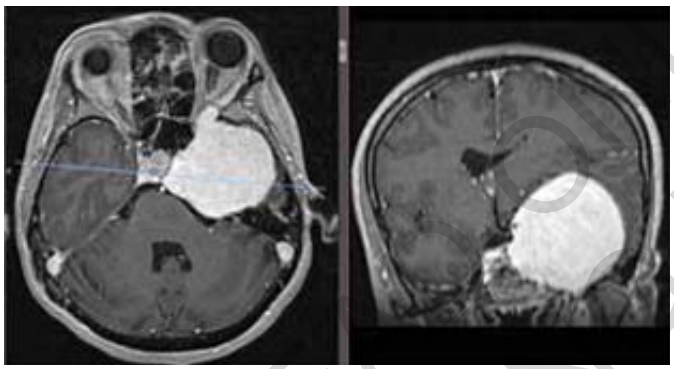

Fig. 1: Pre-operative T1 post-contrast MRI.

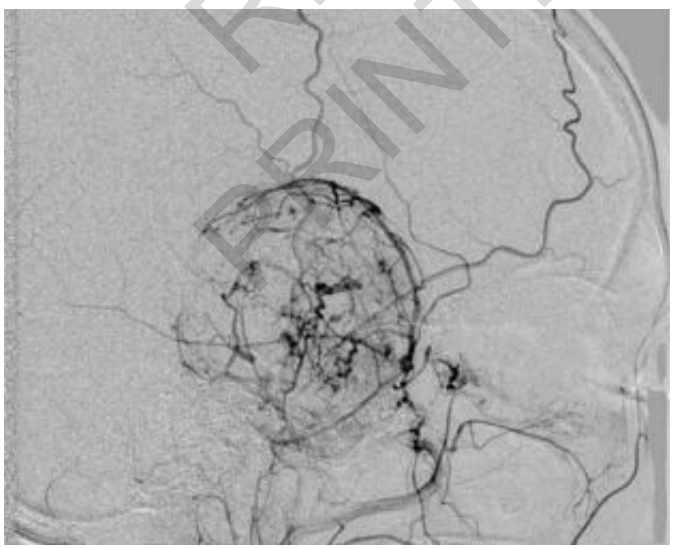

Fig. 2: Pre-operative DSA.

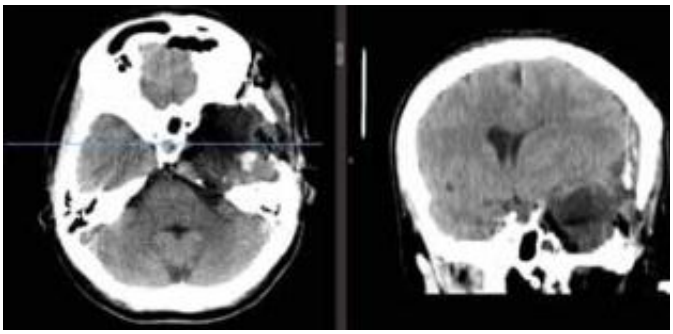

Fig. 3: Immediate post-operative CT-scan.

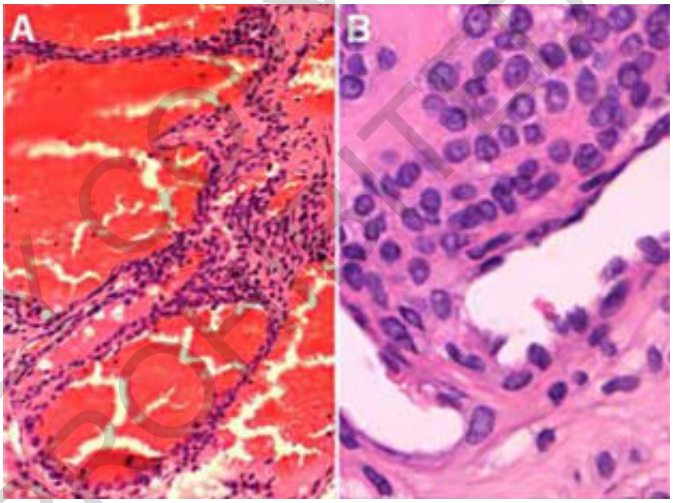

Fig. 4: Dilated blood vessels (A) surrounded by rows and small clusters of small, uniform, round tumor cells (B). A, B, Hematoxylin and eosin, original magnification: A, x 200; B, x 400

Postoperative neurological examination showed a clear improvement in left eye visual acuity; conversely, oculomotor nerve deficit remained unchanged. As for the left ptosis, patient was submitted to a dynamic plastic to take advantage of the improvement in visual acuity in the eye.
Histopathological analysis concluded for glomangioma diagnosis (Fig. 4); in sight of this, we planned radiotherapy treatment of the residual lesion into cavernous sinus. MRI after irradiation was performed 3 months later (Fig. 5).
An annual radiological follow up demonstrated no evidence of tumor recurrence up to the last follow up 7 years after the procedure (Fig. 6).

Discussion

Soft-tissue glomus tumors are benign lesion derived from neuromyoarterial plexus: modified smooth muscle cells of the glomus body, which are specialized arteriovenous

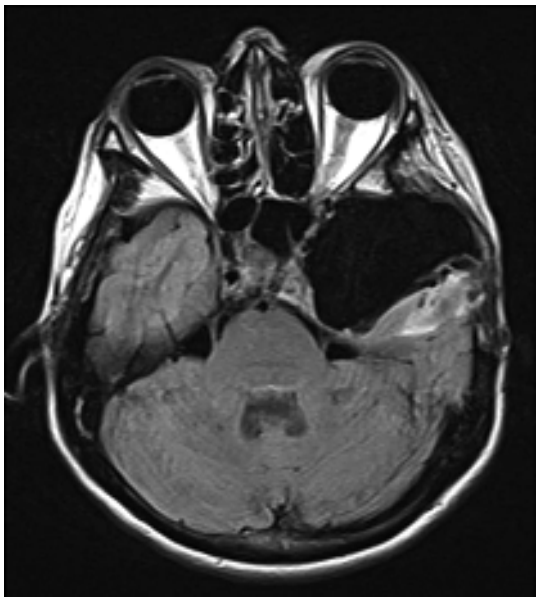

Fig. 5: 3 months post-op control MRI (after radiation therapy).

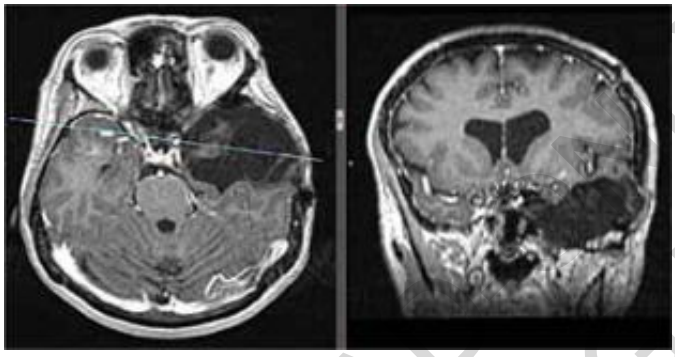

Fig. 6: 7 years post-op control MRI.

shunt that serve a termoregulatory function and are concentrated in the distal extremity dermis. They are best thought of as harmatomas rather than true tumours. There are two main components on microscopy: branching vascular channels and aggregates of specialised glomus cells [1,9].

These lesions most commonly presented as small, elevated, firm nodules that cause exquisite local pain [3].

In addition to the dermis of the distal phalanges, reported sites of soft-tissue glomus tumor include the gastrointestinal and genitourinary tract, bone and nasopharynx [3-10].

These lesions are unrelated to neuroendocrine paraganglioma; marked vascularity on histological examination is the only shared characteristic between the two types of tumor [3].

Our experience was characterized by an uncertain preoperative diagnosis. Magnetic resonance imaging and angiography demonstrated an intracranial but extracerebral lesion with high vascularization.

In literature, there are very few reported cases of glomus tumors with intracranial localization [4,10].

Differential diagnosis with craniofacial schwannomas should be taken into consideration since many reports exists in literaraure [11,12].

To our knowledge, the duration of progression-free survival following treatment was not described.

After surgical and irradiation treatments, our patient underwent to a 7 years follow-up. Imaging studies demonstrated no signs of recurrence. Patient has still a good standard of life.

Conclusion

In this case a subtotal resection followed by radiation therapy determined no recurrence of the disease up to 7 years. To our knowledge, this is the first case described in literature of intracranial soft tissue glomus tumors (glomangioma) in the adult population with a long-term follow-up.

Riassunto

I tumori glomici, o glomangiomi, sono tumori vascolari benigni in genere osservati alle estremità distali degli arti Questi tumori differiscono dai paragangliomi e sono presenti classicamente nella popolazione femminile tra il 4° e il 5° decennio. Localizzazioni intracraniche non sono state descritte in letteratura nella popolazione adulta.

Viene qui presentato il caso di una donna di 32 anni con una storia di 3 mesi di progressiva perdita della vista a sinistra e mal di testa. Una risonanza magnetica preoperatoria ha mostrato una lesione omogenea che si estendeva dal seno cavernoso sinistro alla fossa cranica media inizialmente sospettata di essere un meningioma del seno cavernoso. Alla fine, l'analisi istopatologica si è conclusa per una diagnosi di glomangioma. È stata anche eseguita la RT post-operatoria.

CONCLUSIONI: In base alla nostra esperienza, è molto importante per la gestione clinica considerare i glomangiomi nella diagnosi differenziale di una lesione extra-assiale.

La resezione subtotale seguita dalla radioterapia non ha determinato recidive della malattia fino a 7 anni.

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