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LETTER TO THE EDITOR

Orbital melanocytoma: Multimodal imaging and review of the literature☆

Mélanocytome orbitaire: imagerie multimodale et revue de la littérature

Introduction

Melanocytomas are rare, primary and pigmented tumors. Although melanocytoma were considered as a benign tumor, some reports described a transformation into malignant melanoma [1].

In this case-report, we describe the clinical, radiological and histopathological features of a rare case of an orbital melanocytoma. In addition, the relevant medical literature is reviewed.

Case description

An 11-year-old female, without past medical history, was referred with a two years' history of proptosis of her left eye and a superotemporal eyelid mass. Her best-corrected visual acuity was 20/20 and the clinical examination found a smooth tumor in front of the lacrimal area and leading to an eyeball infero-medial dystopia.

Firstly, a Doppler-ultrasonography was performed and showed a vascular solid lesion. Secondly, a computed tomographic scan demonstrated a large solid hyperdense and well-circumscribed lesion in the right superior orbit, enhanced after intravenous injection of Iobitridol (Guerbet, France) causing a mass effect on the eyeball and repelling the orbital roof without osteolysis (Fig. 1A—B).

Finally, a magnetic resonance imaging (MRI) was performed. The mass appeared hyper intense on T1-weighted images and iso intense on T2-weighted images (with few hypo intense areas) homogenously enhancement after gadolinium injection (gadoteric acid, Dotarem, Guerbet, France) and these results were confirmed by subtraction between T1-weighted images without and gadolinium injection (Fig. 1C—D). The preoperative diagnosis was intraorbital cavernoma.

After multidisciplinary discussion, a surgical treatment with a complete excision was decided (Fig. 1E). There were no postoperative complications.

Gross examination showed a heavily pigmented encapsulated mass measuring 23 mm. Histopathological analysis revealed a proliferation of polyhedral or epithelioid melanocytes (Fig. 1F). Moreover, there were no malignant findings of mitosis or nuclear atypia.

Staining for HMB-45 (antibody identifying melanocytic tumors) was positive whereas staining for S-100 protein and Melan-A protein were negative. The Ki-67 antigen labelling proliferation index was less than 1%. The tumor cells no reacted with CD34 and CD68. The diagnosis of orbital melanocytoma was retained morphologically and on immunohistochemical analysis.

Discussion

Ocular melanocytomas commonly occur near or on the optic disk, they have also been reported to develop in the choroid, ciliary body, iris, conjunctiva or sclera but only a few cases of orbital melanocytoma have been described in the literature [2—9].

Table 1 showed the nine cases of orbital melanocytoma which have been reported in the literature.

The nine orbital melanocytomas reported patient ages ranging from 6 months to 68 years old with a mean patient age at presentation of 34.3 years. These orbital tumors occurred preferentially in men (8/9) and in the right orbit (7/9). Proptosis was the first sign of disease in eight cases and diplopia was present only in two cases. Treatment was surgery in all cases, including complete resection in six cases (66.7%), subtotal resection in two cases (28.6%) and exenteration in one case (14.3%). The two patients with subtotal resection underwent adjuvant therapy (chemotherapy and radiotherapy). Any report has identified malignant transformation or recurrence with the follow-up time ranging from 7 months to 12 years.

On MRI, the tumors appeared hyperintense on T1-weighted images in four cases (57.1%), hypointense in one case (14.3%) and isointense two cases (28.6%). On T2-weighted images, orbital melanocytomas appeared isointense in five cases (71.4%) and hypointense in two cases (28.6%). In all cases, there was a contrast related enhancement after injection of gadolinium. Typically, melanin exhibits high signal on T1-weighted images and low signal on T2-weighted images, which is related to its paramagnetic properties. The varying amounts of melanin within lesions explain these signal variations.

All orbital melanocytomas were positive for HMB-45 and indicated that this tumor derived from melanocytes. In 6 cases (66.7%), immunohistochemical positivity for S-100 protein was found.

☆ *Cas presenté à la Société ophtalmologique plastique reconstructrice esthétique française en mai 2017, Paris, France.*

Table 1 Demographic, clinical and surgico-pathologic findings of the 9 orbital melanocytomas described in the literature.

Author	Age	Sex[b]	Eye	Symptoms	MRI	Treatments	IHC[c]	General association	Recurrence
Tella et al. [2]	35 yo[a]	M	Right	Proptosis	IsoT1−T2, gadolinium +	Subtotal surgical resection + radiotherapy	S100 + HMB45 + chemotherapy	No	No at 5-years of follow-up
Mathai et al. [3]	40 yo	M	Right	Proptosis	−	Total surgical resection	S100 + HMB45 +	No	No
Tsugu et al. [4]	51 yo	M	Right	Proptosis, diplopia	HyperT1−IsoT2, gadolinium +	Total surgical resection	S100 − Melan-A − HMB45 +	Multiple cavernous cerebral angiomas	No at 15-months of follow-up
Sato et al. [5]	49 yo	M	Right	Diplopia	HyperT1−HypoT2, gadolinium +	Subtotal surgical resection + chemotherapy	S100 + HMB45 +	No	No at 12-years of follow-up
Bajaj et al. [6]	6 months	M	Right	Proptosis	−	Exenteration	S100 + HMB45 +	Multiple cutaneous melanocytic nevi and oculodermal melanosis	No at 3-years of follow-up
Tergnago et al. [7]	28 yo	M	Right	Proptosis	HyperT1−IsoT2, gadolinium +	Total surgical resection	S100 + Melan-A + HmB45 +	Ipsilateral Nevus of Ota	No at 7-months of follow-up
Ortiz et al. [8]	68 yo	M	Left	Proptosis, headache, vision loss	IsoT1−T2, gadolinium +	Total surgical resection	S100 + Melan-A + HmB45 +	No	No at 3-years of follow-up
Palicelli et al. [9]	26 yo	M	Right	Proptosis, ocular palsy	HypoT1−T2, gadolinium +	Total surgical resection	HMB45 +	No	No at 3-years of follow-up
Saunier et al.	11 yo	F	Left	Proptosis	HyperT1−IsoT2, gadolinium +	Total surgical resection	S100 − Melan-A − HMB45 +	No	No at 1-year of follow-up

[a] yo: years old.
[b] M: Male; F: Female.
[c] IHC: ImmunoHistoChemistry.