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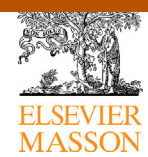

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What is your diagnosis?

Diagnostic trap in relation to an intranasal tumour

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1. Clinical history

Mr. Ra. . . 67-years-old, with no notable history, was urgently referred for facial cellulitis that had been progressing over a period of several weeks in an afebrile context. Clinical examination revealed left hemifacial inflammatory oedema with a lower eyelid fistula and complete ophthalmoplegia and chemosis. The patient reported chronic epistaxis and left nasal obstruction. Nasal endoscopy demonstrated complete filling of the left nasal cavity by a mass lying flush with the nostril and presenting contact bleeding.

2. Questions

Q1: CT scan of the sinuses, followed by facial MRI were requested (Fig. 1). What did they show?

Q2: What management would you have proposed, bearing in mind that the laboratory work-up demonstrated an intense inflammatory syndrome?

Q3: Several biopsies were performed, but histological examination proved to be non-contributive. What management would you propose?

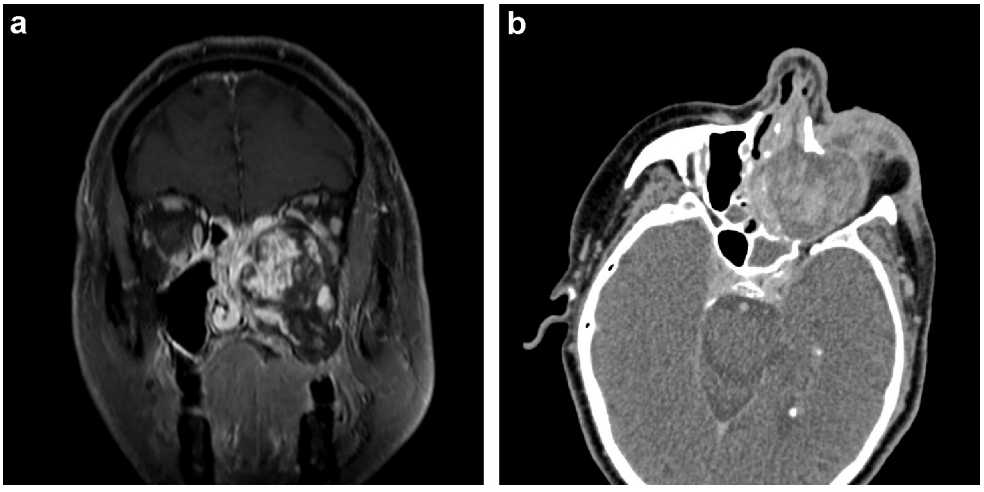

Fig. 1. Gadolinium-enhanced MRI coronal slice and CT axial slice of the patient's lesion on admission. All criteria of malignancy are present on the imaging: bone destruction, extensive nature, heterogeneous contrast enhancement, orbital invasion.

What is your diagnosis?

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3. Answers

Q1: CT scan of the sinuses showed left pansinus disease, centered on the ostiomeatal region, suggesting a destructive (lamina papyracea, posterior, medial and anterior walls of the maxillary sinus), and extensive tumour (intraorbital extraocular extension displacing the medial and inferior rectus muscles; palpebral and malar subcutaneous infiltration) with heterogeneous contrast enhancement.

The appearance of the lesion on gadolinium-enhanced MRI was in favour of lymphoma or carcinoma in view of its enhancement characteristics and its appearance on diffusion-weighted imaging: heterogeneous appearance on T1- and T2-weighted sequences, with slow and heterogeneous gadolinium enhancement, with a high-intensity signal on diffusion-weighted imaging and a markedly decreased ADC (apparent diffusion coefficient).

Q2: A series of intranasal biopsies was urgently performed under general anaesthesia, while the patient was in hospital, for bacteriological, mycological, parasitological and histological examinations. Empirical intravenous antibiotic therapy (ceftazidime and ciprofloxacin) and corticosteroid therapy were instituted without waiting for the biopsy results due to the presence of signs of compressive optic neuritis. Antibiotic therapy was continued for 6 weeks before recovery of normal clinical examination, ophthalmological assessment and laboratory assessment.

During hospitalisation, a second series of biopsies under general anaesthesia had to be performed, as the first series of biopsies was non-contributive. Finally, none of the biopsies performed was able to provide an aetiological diagnosis. Despite the large number of samples, the histology report indicated: “bloody, fibrinoleukocytic material suggestive of an inflammatory nodule with no histological signs of malignancy” and samples for bacteriological, mycological and parasitological examinations remained negative.

Q3: In the presence of these clinical and radiological features highly suggestive of malignancy, while good quality biopsies performed under general anaesthesia were non-contributive, it was decided to perform surgical exploration of the lesion via a combined endonasal and intraoral vestibular approach. Preoperative arteriography was performed for embolization of the internal maxillary artery due to the extremely haemorrhagic nature of the lesion during biopsies. The surgical procedure allowed complete resection of a lesion with a thick capsule, easily dissected from the floor of the orbit and the walls of the maxillary sinus. Histological examination again concluded on nonspecific inflammatory material despite the large quantity of tissue examined (tumour, middle and inferior turbinates, ethmoidal mucosa, anterior and medial walls of the maxillary sinus). A diagnosis of inflammatory pseudotumour was finally adopted.

No recurrence was observed at the 6th postoperative months nor at 1 and 2 years.

4. Discussion

The diagnosis adopted in this patient was that of inflammatory pseudotumour (IPT). This imprecise and nonspecific term corresponds to a lesion of unknown origin. Other terms used in the literature include plasma cell pseudogranuloma, myofibroblastoma, xanthogranuloma, histiocytoma or myofibroblastic inflammatory tumour.

These tumours are rare [1,2], with a ubiquitous distribution, but are usually described in the lungs, liver or spleen in the form of suspicious nodules [2]. Head and neck IPT is fairly uncommon and paranasal sinus IPT is even rarer, as only about thirty cases have been described in the literature [1,3,4]. The orbit is the most common site of IPT in the face [5].

IPT has a worrying clinical presentation, as it always mimics a tumour, as these unilateral lesions are invasive and destructive and associated with severe symptoms: intense pain, cranial nerve paralyses, cervical lymphadenopathy, epistaxis, facial cellulitis, etc. [1,3,5]. CT or MRI imaging reveals signs of local invasion: erosion and destruction of bony walls, extension beyond anatomical limits, contrast enhancement inside the mass [5]. The radiologist very often proposes a diagnosis of lymphoma or carcinoma and recommends urgent biopsy.

However, a key issue related to these lesions is that repeated biopsies are commonly performed due to the nonspecific histological results because the otorhinolaryngologist wants to definitively exclude a malignant disease. Deeper and deeper biopsies are therefore performed, usually under general anaesthesia in order to improve their sensitivity.

The pathogenesis of IPT remains unclear. It appears to be a xanthogranulomatous type of inflammatory reaction secondary to an unknown initial triggering factor. The hypothesis of an auto-immune process or concomitant atypical mycobacteria or actinomyces infection has been proposed, but with no formal evidence [1,3,4].

Histological examination reveals two types of nonspecific features: myofibroblasts and an inflammatory infiltrate (lymphocytes, plasma cells and granulocytes) but no neoplastic features (small number of mitotic figures, polyclonal plasma cell infiltrate, absence of cell necrosis).

Treatment has not been clearly defined and depends on the clinical features [4]. In limited vision-threatening forms, long-term corticosteroid therapy, sometimes initiated at high doses, has been shown to give good results [4,5].

In lesions of the paranasal sinuses, surgery is the preferred treatment [1,4]. The whole operative specimen is sent for histological examination, in order to formally exclude a malignant lesion. Fragments of bony walls must also be sent for histological, bacteriological and mycological examinations.

The literature reports recurrences in 5 to 37% of cases, all sites combined, without specifying the time to recurrence, thereby justifying long-term follow-up [1].

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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