The Journal of Laryngology & Otology
July 2003, Vol. **117,** pp. 564–565

Lymphangioma of the sphenoid sinus

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Abstract
Lymphangiomas are rare benign lymphatic tumours found predominantly in the head and neck region. A case of a cavernous lymphangioma isolated to the sphenoid sinus is described. The authors emphasize the extreme rarity of the isolated sphenoid lymphangioma, as to their knowledge this is the first report in the English literature.

Key words: Sphenoid Sinus; Lymphangioma; Immunohistochemistry

Introduction

Lymphangiomas are rare congenital benign tumours of the lymphatic system that are considered to develop from sequestered elements of primitive lymphatic sacs.[1] The vast majority of them is localized to the head and neck region.[2] Other possible locations are the axilla and the mediastinum.[1] Ninety per cent of cases are detected by the age of two years. According to Batsakis' histopathological classification[3] lymphangiomas can be divided into three groups: (a) lymphangioma simplex or capillary lymphangioma, consisting of thin-walled capillary-sized lymphatic vessels; (2) cavernous lymphangioma, composed of dilated lymphatic spaces; (3) cystic lymphangioma or cystic hygroma which contains cysts of various diameters from a few millimetres to several centimetres. The cavernous type is the most common, over two thirds of all lymphangiomas belong to this group. Their significance is based on the compression of neighbouring normal structures. Its therapy is complete surgical excision which may be difficult due to the infiltrative nature of lymphangiomas.[4]

Case report

A 16-year-old female was referred to our clinic with headache localizing to the frontal region and vertex for five months and paroxysmal dizziness. In her longer-term history there was a purulent meningitis at the age of three months. Neurological, otoneurological and ophthalmological examination and the paranasal sinus radiograph did not reveal any special causes of the headache. Apart from a mild nasal septal deviation her physical status was negative. Computed tomography (CT) scan in the coronal plane showed a soft tissue mass partially involving the sphenoid sinus of normal size (Figure 1). All other paranasal sinuses were negative. With a clinical diagnosis of a sphenoid sinusitis the posterior pole of the left middle turbinate was resected under endoscopic control and then an endoscopic sphenoidotomy was performed and the mass that seemed to be a mucosal swelling of the sinus was completely removed. In contradiction to a haemangioma the colour of the lesion was not different from that of the normal surrounding mucosa. The histopathological examination of the excised tissue revealed a cavernous lymphangioma (Figure 2). The vessel diameters were more variable at the periphery of the lesion. There was only scanty eosinophilic precipitate intravascularly but there was no evidence of red blood cells. After the immunohistochemical processing performed with CD 31 specific monoclonal antibody JC 70 (DAKO, code: MO 823) the endothelial lining of vessels showed a strong CD31 positivity. This confirmed the vascular origin of the tumour. Some days after surgery the patient's headache completely ceased. Now six months after surgery she is disease-free at endoscopic examination of the sphenoid sinus.

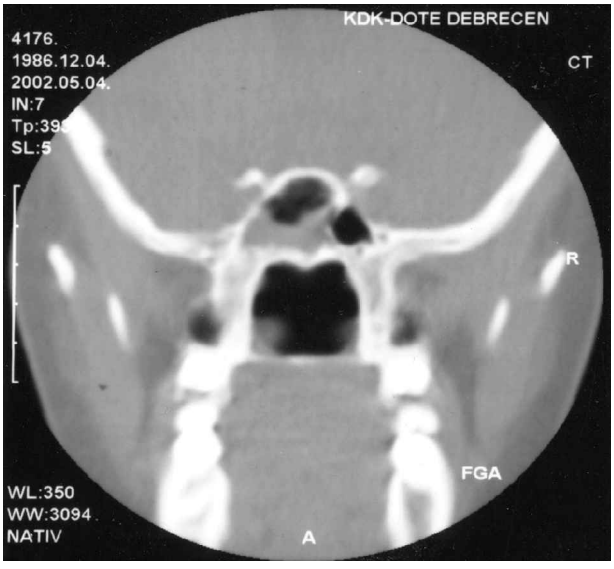

Fig. 1

CT scan of the paranasal sinuses in the coronal plane showing a soft tissue mass in the left sphenoid sinus.

Discussion

Isolated sphenoid sinus disease (ISSD) is a relatively uncommon entity. In a retrospective analysis of 132 cases of ISSD reported by Lawson *et al.* inflammatory disease

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Accepted for publication: 20 March 2003.

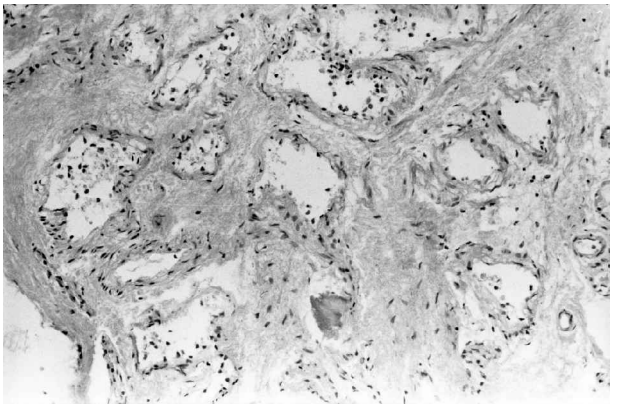

FIG. 2
Cavernous lymphangioma composed of dilated lymph vessels lined by endothelial cells (H&E; ×200).

proved to be by far the most common cause (80 cases).[5] They found 15 malignant tumours. In this group adenoid cystic carcinoma and squamous cell carcinoma proved to be relatively the most common types. There are anecdotal reports of mucoepidermoid carcinoma, primary melanoma or other histopathological types.[5,6] Metastases are also extremely rare, in this group renal cell carcinoma was the most common and there are some reports about the metastases of gastrointestinal, breast, thyroid and hepatocellular carcinomas.[7,8] In some cases adjacent tumours spread into the sphenoid sinus.[5] Lawson *et al.* found 10 benign tumours of which three cases were inverting papillomas.[5] In another report of 122 cases of ISSD the authors found four inverting papillomas.[9] There are scattered reports of schwannomas and plasmacytomas as well.[10] To our knowledge, in the English literature our report is the first of a lymphangioma as an ISSD. In the German literature Schmitt *et al.* reported a 58-year-old woman suffering from a slowly growing sphenoidal lymphangioma.[11] Drut *et al.* reported a paediatric case involving the paranasal sinuses and another case of a mastoid lymphangioma causing massive osteolysis of skull bones.[12] In this report, the tumour was not confined solely to the sphenoid sinus, it involved the frontal sinus and the ethmoid cells, too.

The most common symptoms of sphenoidal tumours are headache, visual loss and cranial nerve palsies (the abducent, the trigeminal and the oculomotorius nerve are involved in most cases).[5] These symptoms can be explained with the vicinity of vital structures to the sphenoid sinus: the sinus cavernosus containing cranial nerves III, IV, V/1, V2, VI, the vidian nerve and the internal carotid artery; the sphenopalatine ganglion, the optic nerve and the hypophysis. The thickness of the bone of the lateral wall is usually less than 0.5 mm and sometimes this wall is totally dehiscent.[13] After extending to the nasal cavity, facial or dental pain, obstruction and epistaxis can also occur. In our case headache was the sole complaint of the patient, which can be explained by the limited extent of the tumour confining to the sphenoid sinus. In the CT scan it is obvious that the bony walls of the sinus are intact. The appearance of the tumour in the CT scan is characteristic of a type I finding according to the classification recommended by Lawson *et al.*[5] In this group the bony borders are preserved, and in the lumen one can find either an air-fluid level or a total opacity or a polypoidal mass. Because of the integrity of the bony walls in our case we did not suggest a magnetic resonance image (MRI) examination and due to the isolated disease we considered the endoscopic approach to be enough for removing the mass.

The histopathological finding showed a cavernous lymphangioma. Transmigration protein CD 31 (also known as platelet/endothelial cell adhesion molecule 1; PECAM-1) is crucial to leukocyte transmigration through intercellular junctions of microvascular endothelial cells.[14] Monoclonal antibodies JC70 and EN-4 are widely used in the detection of fixation-resistant epitopes of CD31: the positivity for this protein confirms the vascular nature of a tumour.[15] This is the reason why we applied the CD31 specific immunohistochemical reaction for the microscopical diagnosis.

- **Cavernous lymphangioma of the sphenoid sinus is a rarity**
- **The mass was removed by endoscopic sphenoidotomy**

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T. Major M.D., takes responsibility for the integrity of the content of the paper. Competing interests: None declared