Glomangioma of the nasal cavity and paranasal sinuses*

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SUMMARY

The glomangioma, a benign vascular tumour, derived from the cutaneous glomus bodies, should not be confused with paragangliomas, which are occasionally also referred to as glomus tumours. Up to now, only eleven cases of a glomangioma of the nasal cavity and the paranasal sinuses have been published. We report the case of a patient with a glomangioma of the ethmoidal aircell system. The tumour was completely removed under endoscopic-microscopic vision via endonasal access. A tumour recurrence was not observed over a period of eighteen months. The clinical signs, diagnosis, therapy and histological features will be discussed with reference to the literature.

Key words: glomangioma, glomus tumour, hemangiopericytoma, nasal cavity, paranasal sinuses

INTRODUCTION

The glomangioma (true glomus tumour) belongs to the group of glomus tumours. They represent benign vascular tumours growing mainly in the skin and in subcutaneous fatty tissue. They derive from modified muscle cells of special arteriolovenular anastomoses, so-called cutaneous glomus bodies (Enzinger and Weiss, 1996).

Glomangiomas should not be confused with chromaffin or non-chromaffin paragangliomas, e.g. chemodectomas, which are often also referred to as glomus tumours (glomus caroticum, glomus jugulare).

According to the distribution pattern of the glomus organs, the most common locations for glomangiomas are the hands and feet (mainly the nailbeds) (McKee, 1996). The mucous membrane of the nasal cavity and the paranasal sinuses are very rarely affected. Only eleven published cases have been found in the literature (Table 1).

CASE REPORT

A 66-year-old female patient complained of recurring nasal haemorrhage and right-hand nasal obstruction for two and a half years. There were no cephalgies or disorders of smell.

Otorhinolaryngology-examination showed a large nasal polypous tumour in the right middle nasal meatus and a deviation of the nasal septum. The CT-scan of the paranasal sinuses revealed an opacity of the right ethmoidal aircell system and the sphenoid sinus. The maxillary and frontal sinuses were inconspicuous, and osseous destructions were not detected (Figure 1).

Septoplasty was subsequently performed in combination with a bilateral conchotomy and an endoscopic-microscopically assisted paranasal sinus operation on the right with endotracheal

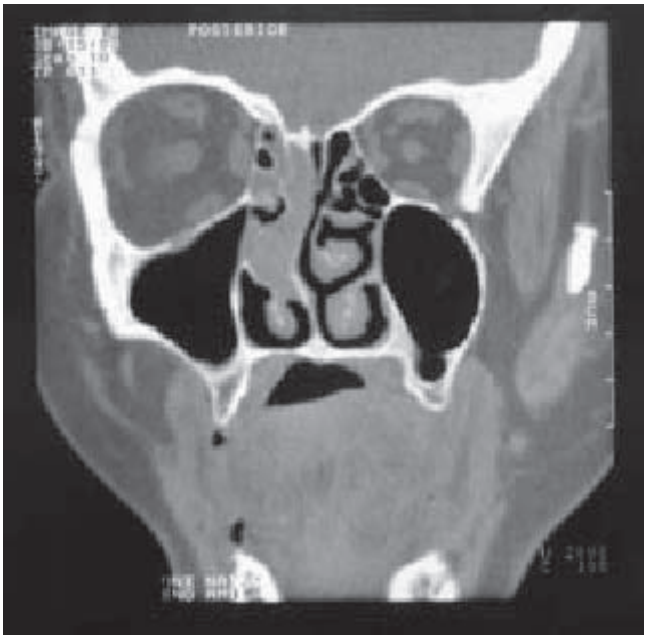

Figure 1. Coronal CT of the paranasal sinuses showing the tumour in the right middle nasal meatus and the ethmoidal aircell system.

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Table 1. Glomangiomas of the nasal cavity and the paranasal sinuses (eleven cases).

	age	sex	localization	complaints	surgical access	period of observation
Pantazopoulos, 1965	45	female	inferior turbinate	nasal obstruction, local pain, bleeding	lateral rhinotomy	8 months without recurrence
DeBord, 1972	33	female	choana	nasal obstruction	endonasal	3 years without recurrence
Fu et al., 1974	71	female	nasal septum	none	endonasal	7 years without recurrence
Fleury et al., 1979	24	male	nasal septum	nasal obstruction, local pain	endonasal	not mentioned
Potter et al., 1984	81	female	nasal septum	none	endonasal	not mentioned
Morais et al., 1986	66	male	nasal vestibule	none	endonasal	not mentioned
Alarcos-L. et al., 1992	55	male	nasal cavity, choana, ethmoidal aircells	nasal obstruction	endonasal	1 recurrence of tumor after 8 months
Hayes et al., 1993	32	female	nasal vestibule	nasal obstruction	endonasal	6 recurrences of tumor within 14 years
Devouassoux-S. et al., 1996	81	female	nasal cavity, sphenoid sinus, ethmoidal aircells	nasal obstruction, bleeding	endonasal	10 months without recurrence
Arens et al., 1997	40	male	inferior turbinate	bleeding	endonasal	3 years without recurrence
Matschiner et al., 1999	9	female	nasal septum	local pain, bleeding	endonasal	not mentioned
	36	female	nasal septum	local pain, bleeding	endonasal	not mentioned
	74	female	nasal septum	nasal obstruction, local pain, bleeding	endonasal	not mentioned

anaesthetisation. While removing the ethmoidal aircell system a reddish, slightly bleeding tumour was discovered and totally excised.

Histologically, the paranasal mucous membrane showed several cavernous, thin-walled blood vessels surrounded by a single or by multiple layers of uniform oval cells. These cells showed round and centred nuclei, a slightly eosinophilic and moderately wide cytoplasm margin and distinct cell borders. They were surrounded by a network of reticular fibers and embedded in loosely fibrotic connective tissue. Cytological anomalies or atypical mitoses were not detectable (Figure 2).

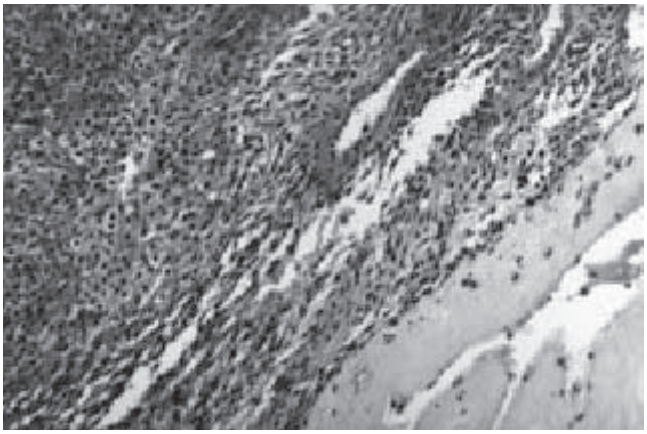

Figure 2. Hematoxylin-eosin-coloring, 1:100
First slice of the glomangioma with cavernous blood vessel (right) and uniform oval glomus cells (left).

Histological inspection pointed to a glomangioma. This diagnosis was confirmed by specific immunohistochemical tests. The tumour cells were alpha-actin- and vimentin-positive. Tests with antibodies against cytokeratins (AE 1/3, KL 1), S 100, HMB 45 and epithelial membrane antigens proved negative. Moreover, the tumour cells were gamma-enolase-, neurofila-

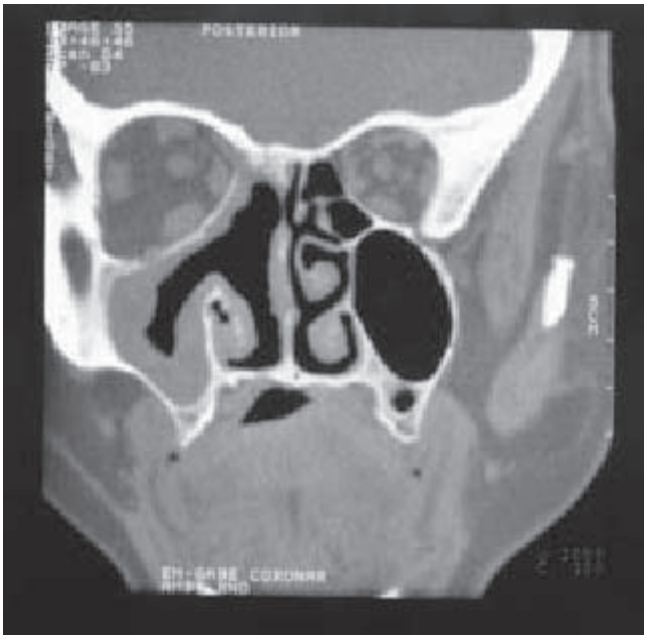

Figure 3. Coronal CT of the paranasal sinuses after total surgical removal of the tumour.

ment- and chromogranin-negative. When testing antibodies against Mib-1, a proliferation activity of tumour cells of less than five percent was found.

A follow-up CT-scan six months after surgery showed no signs of tumour recurrence. Only a marginal swelling of the mucous membrane of the right maxillary and ethmoidal sinuses was noted (Figure 3).

DISCUSSION

Histologically, glomus tumours consist of glomus cells, blood vessels and unstriated muscle cells. Depending on the composition of these components one differentiates solid glomus tumours (25 % of cases), glomangiomas (60% of cases) and glomangiomyomas (15% of cases) (Enzinger and Weiss, 1996; Calonje and Wilson-Jones, 1997).

Glomangiomas are benign tumours which were first described by Wood in 1812. They originate from modified muscle cells of the so-called glomus body, a special form of an arteriolovenular anastomosis (Enzinger and Weiss, 1996; Calonje and Wilson-Jones, 1997). The glomus body is supplied by an arteriole which branches into four small arterioles. These arterioles merge into a thick-walled segment with an irregular lumen, the so-called Sucquet-Hoyer channel. It is limited by a layer of muscle fibers with interspersed epitheloid glomus cells. This channel drains into a thin-walled collecting vein which plays an important role in thermoregulation (Enzinger and Weiss, 1996).

Glomus tumours are relatively rare tumours, with the majority of cases occurring in the forth decade. There is no sex prevalence (Enzinger and Weiss, 1996; Calonje and Wilson-Jones, 1997). They are solitary tumours most frequently localized in deeper layers of the dermis and subcutis of the palms and the nailbeds. These tumours are extremely rarely found in the mucous membranes of the nasal cavity and paranasal sinuses, regions which normally do not contain any glomus bodies. Glomus tumours of this localization are probably derived from pluripotent mesenchymal cells, which differentiate towards glomus cells (Enzinger and Weiss, 1996; Calonje and Wilson-Jones, 1997).

Multiple glomus tumours are found in less than ten percent of the cases. These tumours occur above all in children and adolescents (Wood and Dimmick, 1997). A dominant autosomal inheritance with incomplete penetrance is assumed, as a family history and an association with other malformations are often observed for these tumours (Enzinger and Weiss, 1996).

Macroscopically, a glomangioma manifests itself as a soft purple tumour several millimetres in size which tends to bleed when touched.

This tumour can cause paroxysmal pain which is triggered or aggravated by temperature changes (above all by cold) and mechanical factors (particularly pressure) (Enzinger and Weiss, 1996; McKee, 1996; Calonje and Wilson-Jones, 1997). In addition, glomangiomas of the nasal and paranasal region become symptomatic by recurrent haemorrhage and gradually progressive nasal obstruction. Therefore, they are often diagnosed at an early stage before adjacent tissue structures are destroyed.

The most important differential diagnosis is the hemangiopericytoma, a relatively rare tumour first described in 1942 by Stout and Murray. Hemangiopericytomas occur in middle-aged and elderly patients. Preferred localizations are the deep soft tissues of the lower limbs, pelvis and retroperitoneal space (Enzinger and Weiss, 1996; McKee, 1996; Calonje and Wilson-Jones, 1997). These tumours are rarely found in the nasal cavity and paranasal sinuses, the meninges or the orbita (Enzinger and Weiss, 1996).

Histologically densely packed, short fusiform cells with oval nuclei are observed. They possess small cytoplasm margins and diffuse cell borders. These cells are grouped around numerous thin-walled ramifying blood vessels. The hemangiopericytoma was originally considered to derive from pericytes because of this growth pattern.

But tumours with a hemangiopericytoma-like growth pattern often show a non-pericytic differentiation with regard to immunohistology and ultrastructure. Rather, they tend to express markers found in tumours of a different histogenesis, as for example synovial sarcomas, mesenchymal chondrosarcomas, solitary fibrous tumours, or benign fibrous histiocytomas. The majority of these tumours seem to derive from pluripotent stem cells (Enzinger and Weiss, 1996; Schurch et al., 1990).

Hemangiopericytomas are tumours with an uncertain status. An increased cell density, extended necroses and elevated rates of mitosis (exceeding 4/10 HPF) are potential criteria of malignancy and can be accompanied by an increased rate of metastasis as well as an increased recurrence rate (Enzinger and Weiss, 1996; Calonje and Wilson-Jones, 1997).

Glomus tumours are usually benign, so that a complete local excision is sufficient. The recurrence rate is about ten percent. Recurrences are probably the result of incomplete removal. Higher recurrence rates are observed only for so-called infiltrating glomus tumours, which are an extremely rare variation with poorly defined margins (Calonje and Wilson-Jones, 1997; Gould et al., 1990; Hayes et al., 1993).

The case reported by Hayes et al. in which six recurrences occurred within fourteen years in spite of repeated complete surgical removal of the glomus tumour in the nasal vestibule could present such a tumour (Hayes et al., 1993).

The complete surgical removal of tumour is the method of choice for managing histologically diagnosed glomangiomas. Depending on size and localization of the tumour in the nasal and paranasal regions, endo- and extranasal approaches are considered for surgical access (Draf and Berghaus, 1993). If possible, the less invasive endonasal approach should be preferred, which leaves the osseous structures intact. Next to the lower complication rate observed after endonasal approaches the aesthetic aspect -in particular the absence of a facial incision- merits consideration (Iro and Hosemann, 1993).

Radiotherapy merely achieves a containment of tumour growth and should only be employed if the tumour is inoperable (Fleury et al., 1979).

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