Cystic Schwannoma of the Maxillary Sinus: Clinical and Radiological Findings

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Abstract: Schwannomas are common tumors of the nerve sheath that frequently involve the peripheral nerves of the head and neck. This benign slow-growing neoplasm is composed of nerve-supporting cells, and rarely affects the paranasal sinuses. This lesion usually occurs in the middle-aged patients, with an equal sex distribution. Schwannomas are usually solid tumors, although cystic components are rarely seen in head and neck area. MRI can prompt the diagnosis that however relies on histological examination. The present study reports a case of schwannoma of the maxillary sinus, diagnosed in a 38-year-old woman that presented nasal obstruction, pain in the right facial area and hypoesthesia in the area of right infraorbital nerve for past six months. Clinical presentation, radiologic aspects, histological features and treatment of this tumor were reported.

Keywords: Schwannoma, Maxillary sinus, Rare tumors.

1. INTRODUCTION

Schwannoma is benign slow-growing neoplasm composed of nerve-supporting cells that affects rarely the paranasal sinuses [1]. Embryologically it originates from neuroectoderm, and typically arises from the branches of the trigeminal nerve or from autonomic nerves in and around paranasal sinuses [2]. In particular, approximately 45% of these tumors occur in this region [3], and most of them arise along the vestibular portion of the eighth cranial nerve [4]. Only 2% to 4% of schwannomas affect the nasal cavity and the paranasal sinuses [3,5], occurring in the middle-aged patients with an equal sex distribution [6]. In the paranasal sinuses they are reported mainly in the nasal cavity and ethmoid compartment, less frequently in maxillary, sphenoid and frontal sinuses [7]. The reason for this difference in the frequency of occurrence could be the more abundant innervations of these regions compared to other paranasal sinuses [1].

The pathologic features of this tumor is not well documented; in fact, Schwannomas are usually solid tumors, although cystic components are rarely seen in head and neck area [3,8].

The clinical symptoms depend on the size and the location of the tumor [9]. The radiological images show the sinonasal schwannoma like well-demarcated solitary tissue mass, usually attached to or surrounded by nerve and associated to bone remodelling and erosion for the constant pressure of the mass on the surrounding structures [1,7].

The case reported in the present study is of interest due to the relative rarity of the pathology and the difficult differential diagnosis with other pathologies, like polyp and mucocele, the commonest sinonasal affection.

2. CASE REPORT

In May 2011, a 38 years-old woman was referred to Clinic of Dentistry and Maxillofacial Surgery, Policlinico G.B. Rossi, Verona, for pain in the right facial area, a sensation of nasal obstruction and tearing of long duration, in addition to hypoesthesia in the area of the right infraorbital nerve for six months. The symptoms were diagnosed as a right maxillary sinusitis. The CT was not available, due to the patient’s refusal to undergo this diagnostic procedure, but MRI was undertaken to further analysis of the lesion and it revealed the presence of a mass, markedly high signal on T2-weighted sequences, indicating a partially liquid component, evenly distributed, that occupied the right maxillary sinus (Figures 1 and 2). The MRI provided evidence of a lesion in the posterior wall of the right maxillary sinus, whose extension reaching up to the posterior part of the infratemporal fossa. After intravenous infusion of paramagnetic contrast agent, the walls of the lesion showed thickening and vascularisation, while the post-
erior part revealed the presence of a small solid component. Compression and thinning of the orbital wall were identified, without evidence of infiltration in the right orbit. A diagnosis of capsulated neoplasm was considered, with evidence of slow growth and remodelling of the surrounding bony structures. The lesion appears like benign in origin and mildly aggressive. The patient underwent a surgical excision of the mass through Caldwell-Luc approach. After a sublabial incision on the right maxillary gingiva, soft tissue and periostium were elevated, and the anterior wall of the right maxillary sinus was open. Surgery confirmed the space-occupying lesion and the absence of infiltration of the adjacent bones. After the complete removal of the tumor, histological examination was performed. Histopathology of the tissue sample revealed a mesenchymal neoplasm with fused cells without evidence of mitotic activity. The tumor capsule was confirmed as being intact. The immunohistochemical examination was positive for antibodies S100, NSE and CD57, giving a definitive diagnosis of schwannoma. The proliferative activity evaluated with Ki-67 antibody was lower than 10%, and therefore the neoplasm was labelled as benign (Figure 3). The postoperative course was uneventful, and there was no recurrence during the follow-up period. After 12 months, the patient drop out the follow-up.

Figure 1: MR T2-weighted images acquired on the coronal [fast spino echo Tr 3300, Te 96.8, echo train length 10] plane after intra-venous paramagnetic contrast show a cystic expansive lesion in the right maxillary sinus: a solid component of the cystic wall is better seen enhancing after contrast along the posterior border of the lesion.

Figure 2: MR T2-weighted images acquired on the axial plane after intravenous paramagnetic contrast.

Figure 3: Histological examination of tissue sample: benign schwannoma of maxillary sinus with spindle cells and distinct Antoni A type areas (hematoxylin and eosin, x200).

3. DISCUSSION

Schwannomas of head and neck region are typically described as solitary encapsulated lesions that arise in the first or second division of the trigeminal nerve. On the contrary, when these tumors are multiple, they are typically found in Neurofibromatosis type 1, a disease associated to mutation at the NF1 gene [4]. Another form of neurofibromatosis was recently recognized,
called schwannomatosis, and a case of bilateral maxillary sinus schwannomas in a patient with this disease was reported very recently in literature [10].

Masses that occupy the paranasal sinuses cause symptoms such as nasal obstruction, epistaxis, rhinorrhea, anosmia, and sometimes headache, sensation of facial swelling and pain. Exophthalmos, epiphora and progressive visus reduction are less frequently occur, mainly related to the involvement of orbital region [11]. In the present case, the long-term symptoms, such as the pain in the facial area, the nasal obstruction and tearing, and the right infraorbital nerve hypoesthesia are in agreement with those reported in the literature [8,12,13]. In fact, schwannomas are slow-growing tumors that can reach a large size before they become symptomatic, and facial pain was reported as the most common symptom with maxillary sinus lesions [14]. The duration of symptoms before diagnosis is very wide, ranging from 1 to 48 months [1]. In the presence of a fluid-filled lesion in a paranasal sinus, in addition with a compatible clinical history of sinusopathy, the suspected diagnosis is a chronic inflammatory process or a mucocele [15]: considering the limited literature currently available on the subject, it is very difficult to consider schwannomas amongst the differential diagnoses due to its cystic component [16]. In comparison to mucocele, schwannoma has an irregular thickness of the lesion walls, as shown in this patient, and the presence of solid material projecting into the central cystic area. CT scan or MRI can evaluate the presence of thickening and vascularisation of the wall and the presence of small solid component. On the contrary, mucoceles show no enhancement [3]. Only investigation with an MRI with contrast agent allowed identification of the neoplastic component of the cystic lesion. Moreover, differential diagnosis with rare malignant peripheral nerve sheath tumors is necessary. A contrast-enhanced CT scan can evaluate the vascularisation of the lesion, but MRI with gadolinium is investigation of choice for identification and examination of the suspected nerve lesions of sinonasal cavity [4]. On an MRI scan, schwannomas appears with an intermediate signal intensity on T1-weighted images, whereas a T2-weighted signal varies depending on the tumor structure [17]. Therefore the correct diagnosis of schwannoma was only confirmed by histopathological analysis of the surgical sample.

Macroscopically, sinonasal schwannoma range in size up to 7 cm and the cut surface show tan-grey, yellowish, solid to myxoid and cystic tissue, usually with haemorrhage [6]. Microscopically, this tumor is characterized by two pattern, originally described by Antoni almost a century ago, called “Antoni A” and “Antoni B” [18]. The present case showed a prevalence of Antoni A areas, that consisted in hypercellular areas with fasciculated pattern, characterized by spindle cells with elongated nuclei arranged in waves, drifts and whorls. Instead the Antoni B areas are hypocellular areas with reticular pattern, made up of loose tissue, lacking the arrangement in the bundle and palisades [19]. Typical is positivity for S100, even if these results do not differentiate between a benign and malignant neof ormation [20]. The distinction between benign and malignant schwannoma is evident only with absent or scarce mitotic activity and the lack of necrosis, significant cellular atypia and infiltrative aspects seen with microscopic examination [21]; instead the lack of encapsulation is not a pathognomonic feature of malignancy. Moreover, local bone remodeling without infiltration is typical of schwannoma, particularly in the oro-sinusal region; in fact, in this case an orbital wall thinning was present, without infiltration of the right orbit. As reported by others, this feature is not indicative of malignancy [22]. The malignant transformation of schwannomas are very rare; in fact, there are few cases in literature of malignant Schwannomas that arose from long standing benign tumors [23]. A diagnosis of capsulated neoplasm was considered, with evidence of slow growth and remodelling of the surrounding bony structures.

Kim et al. [1] speculated that the rarity of cystic degeneration of schwannomas in paranasal sinuses is due to earlier detection of these tumors. In fact, as reported in the literature, cystic degeneration is more frequent in larger schwannomas, due to insufficient blood supply [24].

Surgical excision is the elective treatment of schwannoma. Some authors describe the problem in choosing of the best surgical approach and underline the diagnostic and therapeutic importance of sinonasal endoscopy [2]. In fact, this procedure allows to perform a preoperative biopsy, an accurate preoperative assessment of the tumor extension, and the complete excision of the mass.

Important factors that influence the choice of surgery are the size of the schwannoma and its location in the oro-sinusal region. Endonasal endoscopic approach or a combination of lateral rhinotomy and Caldwell-Luc and external frontoethmoidectomy are used for the excision of schwannomas situated in the nasal cavity or
paranasal sinuses [12,25,26], while for tumors of pterygopalatine fossa, endoscopic transnasal precranial recess-maxillary sinus approach was recently proposed [27].

4. CONCLUSION

The differential diagnosis of sinonasal schwannoma is difficult, due to the lack of specificity of the symptoms and the rarity of the localization. CT and MRI are useful for preoperative diagnosis and surgical planning, even if may sometimes suggest a misleading diagnosis. The present study underlines that cystic schwannoma, although rare, should be kept in mind in the differential diagnosis of cystic masses of the maxilla. The correct diagnosis requires a histopathological analysis, which reveals the two typical patterns of this tumor, called “Antoni A” and “Antoni B”. The treatment of choice for schwannomas of maxillary sinus is surgery, using a conservative approach. In fact, by using endoscopic procedures, the small tumors may be amenable to transnasal endoscopic resection, with an excellent prognosis. In the case presented in this report, an external approach was chosen due to the size of tumor.

A correct preoperative diagnosis of sinonasal schwannoma is important because a complete surgical excision of this lesion is curative, avoiding unnecessary extensive excision, and the risk of recurrences is very low, as well as the risk of malignant transformation.

REFERENCES


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