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Endoscopic treatment of an ethmoidal solitary fibrous tumour

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Abstract
Solitary fibrous tumours are rare neoplasms, which occur most frequently during adulthood at the level of the pleura. Recently, these tumours have been demonstrated in extraserosal sites such as the nasal cavity. A case of a solitary fibrous tumour arising at the level of the ethmoid and presenting in a 54-year-old woman, is reported. The tumour was removed using an endonasal endoscopic approach that permitted the monobloc excision of the lesion. The patient is without evidence of disease 18 months after surgery.

Key words: Head and Neck Neoplasms; Fibrous Tumour, Solitary; Paranasal Sinuses; Nasal Cavity; Endoscopy, surgery

Introduction

Solitary fibrous tumours (SFT) are rare tumours arising mostly at the level of the pleura and less frequently at the level of the other serosal surfaces.[1–3] The histogenesis of SFT is controversial. Some authors have hypothesized a mesothelial origin, however, many reports assert a mesenchymal cell origin with evidence from immunohistochemical and ultrastructural studies.[2,4,5] Recently these tumours have been documented in a variety of sites including the mediastinum,[6] lung,[7] liver,[8] orbit,[9,10] infratemporal fossa[11] and upper respiratory tract, specifically the nasal cavity, paranasal sinuses and nasopharynx.[4,5,10–15] This tumour has two main histological patterns that are usually present together and to different degrees within the same neoplasm: solid spindle and diffuse sclerosing.[16] The solid pattern is mainly characterized by a cellular component, while the stromal and fibrous pattern may reveal a sclerosing neoplastic component. Up to 23 per cent of pleural SFTs behave aggressively.[2] However unreliable, the aggressive behaviour is thought to be associated with the large size of the lesions, hypercellularity, increased mitosis, pleomorphism, necrosis and haemorrhage.[2] The main treatment for SFT is complete surgical excision, although radiotherapy and chemotherapy have been used for the treatment of incompletely resected lesions.[5,17] In this report a complete endoscopic resection of an SFT of the ethmoid is presented.

Case report

A 54-year-old woman presenting with a one-year history of right nasal obstruction, was referred to our Institution for a nasal endoscopy. The endoscopy revealed a pinkish smooth polypoid mass that completely occupied the right nasal fossa, and which arose apparently at the level of the middle meatus and involved the medial aspect of the middle turbinate and anterior ethmoid. A biopsy was subsequently taken from the lesion, and the histopathological report revealed an SFT; furthermore this spindle cell tumour was positive for vimentin and CD34 staining. The subsequent radiological assessment was accomplished by means of an enhanced computed tomography (CT) scan of the facial skeleton which showed an homogeneously enhancing mass occupying the right nasal fossa, and reaching the roof of the ethmoid without breaching into the anterior cranial fossa (Figure 1(a) and (b)). Therefore, it was decided to remove this lesion using a transnasal endoscopic approach. This approach was characterized by an *en bloc* resection of the tumour accomplished by a complete right sphenoethmoidectomy with an ipsilateral middle turbinectomy. The monobloc dissection was achieved through the elevation and resection of a mucoperiosteal flap surrounding the tumour. The definitive histopathological report revealed an unencapsulated SFT, which had been suspected preoperatively at the time of the biopsy. Histologically a patternless arrangement of plump spindle cells within areas of different cellularity were noted (Figure 2). Moreover, there was no evidence of hypercellularity, increased

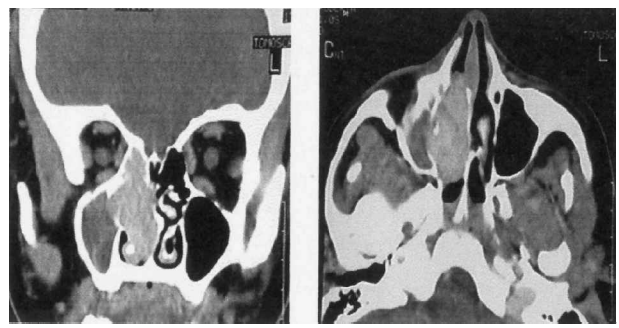

Fig. 1
(a) and (b) Pre-operative coronal and axial view of an enhanced CT scan showing the tumour in the right nasal fossa.

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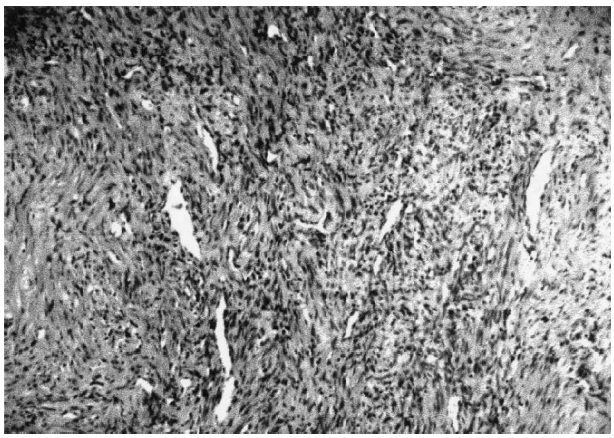

FIG. 2

Photomicrograph of the tumour showing the patternless arrangement of the plump spindle cells.

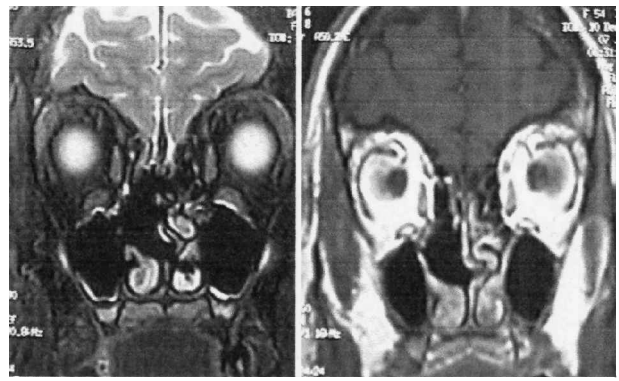

FIG. 3

(a) and (b) Post-operative coronal view of T1- and T2-weighted MRI demonstrating the complete resection of the SFT.

mitosis, pleomorphism or necrosis. Immunohistochemical studies on paraffin-embedded tissue sections showed staining of the tumour cells for vimentin and CD34. There was no immunoreactivity with antibodies to keratin, S-100 protein or desmin. The post-operative course was uneventful and the patient is disease free after 18 months (Figure 3(a) and (b)).

- **Solitary fibrous tumours are rare neoplasms occurring usually in adults at the level of the pleura**
- **Extraserosal sites have been reported including the nasal cavity and paranasal sinuses**
- **Treatment was a monobloc excision using an endonasal endoscopic approach**

Discussion

SFT is a well-recognized entity occurring in the serosal surfaces and most commonly at the level of the pleura.[1–3] The solitary fibrous tumour occurs more frequently in adults of all ages and males seem to be affected as frequently as females.[18] Histologically, these tumours are formed by plump spindle cells arranged in a patternless fashion in a collagenous background. Typically there are hyper- and hypocellular areas and prominent vascularity within the lesion that results in a haemangiopericytoma-like pattern. Immunohistochemical assessment permits the differentiation of the SFT from other fibrous or spindle-cell neoplasms of the upper respiratory tract such as haemangiopericytoma, angiofibroma, fibrous histiocytoma, schwannoma, leiomyoma, fibromatosis and fibrosarcoma.

Generally, poor prognostic factors are the result of large size, hypercellularity, increased mitosis, pleomorphism and necrosis.[2,19] England *et al.* pointed out that the majority of malignant fibrous tumours had mitotic counts of more than four mitotic figures for 10 high-power fields, were larger than 10 cm in diameter, and were haemorrhagic and necrotic.[2] However, not all tumours whether benign or malignant behave as mentioned above, and no reliable guidelines for tumour prognosis have been established.[4,13]

Endoscopic treatment has been successfully employed in the last decade especially for the treatment of benign nasal and paranasal tumours. In general, the endonasal endoscopic approach permits good magnification, visualization, angled vision and illumination when compared to the traditional external approaches. In this report the endoscopic treatment of an SFT arising from the ethmoid was described. The lesion was assessed pre-operatively by means of a clinical endoscopic examination, biopsy and through a radiological assessment by means of an enhanced CT scan. These pre-operative steps were of paramount importance in appropriately assessing the nature and extension of the lesion, in order to choose the correct surgical approach. England and Witkin stated that resectability represents the most important prognostic factor of the SFT.[2,5] In our case, tumoural removal was *en bloc* and it was achieved by means of an endoscopic sphenoethmoidectomy, with ipsilateral middle turbinate resection, through the elevation and subsequent resection of a mucoperiosteal flap surrounding the lesion. The definitive histopathological report confirmed the absence of aggressive characteristics of the SFT such as hypercellularity, increased mitosis, pleomorphism and necrosis. Furthermore the immunohistochemical positivity of the tumour upon staining with vimentin and CD34 helped us to identify this tumour and differentiate it from other lesions of the sino-nasal tract.

Surgery represents the treatment of choice, although radiotherapy and chemotherapy have been used in the treatment of SFT.[17] In fact, Goodlad and Fletcher showed a case of mediastinal SFT which was treated by radiotherapy only and the patient was free from recurrence 19 years later. The recurrence of an SFT has been noted up to 31 years after surgery, thus a long follow up is required for this tumour. Gold *et al.* stated that recurrence and metastases of SFTs depend mostly on the presence of a tumour size greater than 10 cm and on a histological malignant component within the tumour.[18] Owing to the poor outcome the author also suggested an attentive follow up for these high risk patients.[18]

Radiological findings of both magnetic resonance imaging (MRI) and enhanced CT scan have been described for the assessment of SFT.[20–23] The CT findings reported vary from a heterogeneous to a homogeneous strongly enhancing well-defined mass, with areas of calcification and/or necrosis.[21,23] Instead, after MRI, the SFT can be isointense to the muscle on T1-weighted images and hypointense on T2-weighted images, although Shin *et al.* reported that T2-weighted images can be hyperintense especially for those tumours which, at the pathologic examination have hypocellular and collagenous

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sclerotic areas.[20,22] In this report the enhanced CT scan showed a homogeneous enhancing mass without evidence of necrosis and/or calcification.

In conclusion, the endonasal endoscopic approach can be successful in achieving the complete removal of benign tumours such as SFT arising at the level of the sinonasal tract. This type of treatment also has the advantage of no external incision, less blood loss, low morbidity, shorter hospital stay and possible repetition in cases of recurrence when compared to traditional external approaches. Nevertheless, a long follow up for patients treated for SFT of the sinonasal tract is warranted. In fact, a SFT is an exceedingly rare occurrence and the short follow up of the cases involving the nose and paranasal sinuses which have been reported in English literature was far too short to give an adequate indication of a minimum follow-up period for long-term recurrences.

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