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Case Report

A Rare Case of Recurrent Orbital Solitary Fibrous Tumour with Intracranial Extension

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Abstract

A 47-year-old lady, presented with progressive proptosis of left eye with deterioration of vision. She had a history of left solitary fibrous tumour and had undergone left frontal craniotomy and orbitotomy in 2004. Surveillance Magnetic resonance imaging (MRI) six years later showed tumour recurrence with intracranial extension. However, she did not follow-up and only presented again 3 years, later. Tumour resection and left exenteration was performed. Histology showed 'patternless' pattern of neoplastic cells, and CD34 staining was diffusely positive. Diagnosis of recurrent solitary fibrous tumour with intracranial extension was made.

Keywords: CD34, exenteration, orbit, proptosis, solitary fibrous tumour

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Introduction

Solitary fibrous tumour (SFT) is a rare spindle-shaped neoplasm which was initially believed to be arising from mesothelium. Classically, it was localised in the pleura (1). Extrathoracic SFT can be found in the skin, paranasal sinuses, orbit and other sites. Generally, the SFT is a benign tumour with indolent behavior and malignant transformation is uncommon. Previously, SFT of orbit was rarely diagnosed but with presence of CD34 staining, the diagnosis of SFT becomes more common. Here, we report a case of recurrent SFT with intracranial extension.

Case Report

A 47-year-old Chinese lady presented with painless and rapidly progressive left eye proptosis associated with blurring of vision of 2 months duration. She denied any diplopia, or history of recent trauma. Her family members noticed she had behavioral changes and insomnia. However, there were no symptoms of increased intracranial pressure or constitutional symptoms. She had a history of left orbitotomy and craniotomy with excision of tumour for an intraconal tumour 9 years ago. Intraoperatively, the tumour was excised, however parts of the tumour that were adherent to levator palpebrae superioris and medial rectus were diathermised. Histology confirmed the diagnosis of solitary fibrous tumour and immunohistochemical studies showed positivity towards CD34. She was well until recurrence of tumour was detected 6 years post-surgery during tumour surveillance. Magnetic resonance imaging (MRI) revealed an intraconal mass in the medial part of retro-orbital region, measuring 2.2 cm x 2 cm x 2.5cm with evidence of optic nerve compression by the tumour. However, patient defaulted her follow-up as she sought for traditional treatment. She only presented again with the current symptoms 3 years later.

Left visual acuity was 1/60, with presence of left relative afferent pupillary defect (RAPD). External

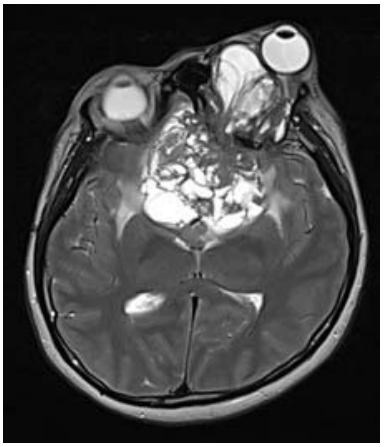

Figure 1: MRI of orbit and brain show T2 weighted image of large extraconal mass pushing the globe inferotemporally and it breaches the cribriform plate to extend intracranially into both frontal lobes.

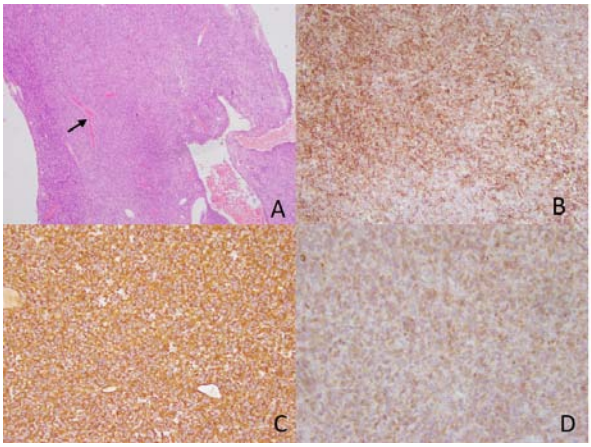

Figure 2: a) H&E (4x) stain shows branching vessels in solitary fibrous tumour (arrow), positivity towards b) CD34 c) CD99 d) BCL-2.

examination showed a left non-axial proptosis, with restriction of extraocular muscles movements in all directions. The proptosis was 8mm on Hertel measurements, and retropulsion test was positive. Intraocular pressure was normal. Left optic disc was swollen with tortuosity of vessels. Right eye showed normal findings. Optic nerve function tests showed the left optic nerve was compromised.

MRI of brain and orbit showed heterogenous enhancing extraconal mass at medial aspect of the orbit measuring 5.6 cm x 5.1 cm x 6.0 cm, exhibiting mixed heterogenous signal intensity on T1 and T2 weighted images with multiseptations and cystic components within. There was clear demarcation between the mass and the left globe, but no clear fat demarcation between the mass and medial rectus, inferior rectus, superior rectus, superior oblique and left lacrimal gland, suggestive of infiltration. The lateral rectus was spared. The left optic nerve was infiltrated by the mass. The mass breached the left lamina papyracea to extend into the ethmoidal sinus, crossing the midline to compress onto contralateral intracanalicular segment of the right optic nerve. The mass breached the cribriform plate and extend intracranially into both frontal lobes resulting in mass effect.

The tumour was accessed via bifrontal craniotomy. Tumour excision and left exenteration was done. Intraoperatively, the recti muscles and lacrimal gland were enlarged. The tumour was believed to be originating from the medial side of the globe, likely from the medial rectus, encasing the optic nerve.

Histopathological examination showed a tumour composed of ‘patternless’ pattern of neoplastic cells exhibiting bland spindle to ovoid nuclei and faintly eosinophilic cytoplasm, with presence of hyalinised collagen in between the neoplastic cells. There was local infiltration of malignant cells into extraocular muscle bundles and involvement of posterior margin. Immunohistochemical staining showed positivity for CD34, CD99, Vimentin and focally positive for SMA and BCL-2. The staining was negative for EMA, desmin, CK, CD31 and S100.

Post-operatively, the patient was assessed by oncology team however there was no role of radiotherapy. Yearly surveillance MRIs did not show any residual and recurrence of the tumour. She is not on any prosthesis. She is currently well, being employed and happy with her life.

Discussion

Solitary fibrous tumour is an indolent benign neoplasm that affects mainly middle-aged adults. In 1931, Klemper and Rabin described it as 'localised mesothelioma', which classically occurs in the pleura (1). SFT of orbit was first described in 1994 by Westra et al. in presence of strong and generalized immunohistochemistry staining with CD34 (2).

Despite SFT being a histological diagnosis, radioimaging plays an important role in diagnosing SFT. Magnetic resonance imaging (MRI) of SFT showed T1 signal intensity that is isointense to gray matter with heterogenous gadolinium enhancement (3). Low T2 signal indicate presence of calcification or a fibrous lesion. In SFT, T2 image shows hypointense to gray matter due to lack of local water density in collagenous stroma in SFT[5]. However, in our patient

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MRI showed heterogenous signal intensity in both T1 and T2 weighted images. The heterogeneity is believed to be due to presence of fibrous stroma intermixed with the prominent vessels (4).

SFT should be differentiated from other soft tissue tumour such as angiomatoid fibrous histiocytoma (AFH) as they share similar histological features. Histological features of SFT show presence of branching hemangiopericytoma-like vessels, while in AFH the blood vessels arrangement is well-formed. With advances in immunohistochemical staining, SFT can be distinguished from AFH. SFT shows diffuse staining with CD34 while AFH shows negative staining towards CD34 (5). 95% of cases of SFT express CD34. About 70% cases with SFT showed positivity with CD99 and 20-30% with EMA (6). Bcl-2 is non-specific, as it is also seen in other mesenchymal neoplasms. SFT shows negative staining with S100, Cytokeratin, EMA, SMA and desmin. Strong positivity of CD34 also indicate benign nature of SFT, as compared in malignant transformation of SFT, the tumour has high mitotic rate with atypia and loss its CD34 immunoreactivity (7).

Incomplete resection of tumour is the most important predictor for local recurrence. Alexandrakis and Johnson reported a case of incomplete excision of SFT in 14-year-old girl lead to recurrence (8). We believe that our patient developed recurrence due to inability to excise the adherent part of the tumour bulk at initial resection. Patient was at risk of developing post-operative diplopia which might affect patient's quality of vision in attempt to excise the adherent part of the tumour. However, metastases in SFT has not yet been reported.

Intracranial extension is uncommon; however the tumour can breach the lamina papyracea to infiltrate into frontal lobes or via the bony foramina of the orbit. There were 3 cases of SFT with intracranial extension reported previously, in which all of the three cases recurred due to incomplete excision of the tumours (9,10,11).

In conclusion, the advances in immunohistochemical staining assist in establishing the diagnosis of SFT and differentiating it from other tumours. Total resection of the tumour is important to prevent recurrence.

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