CASE REPORT

Spindle cell lipoma

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SUMMARY

Spindle cell lipomas (SCLs) are a group of benign lipogenic tumours, typically arising in the posterior neck, upper back and shoulder of elderly male patients. Approximately 80% of these tumours arise in characteristic location, but 20% arise in unusual locations, thereby making these cases more difficult to diagnose. We present a case of SCL occurring in the right periorbital region of a 14-year-old boy. The MRI was suggestive of possible malignancy. Diagnosis of neurofibroma was made on incisional biopsy. However, the histopathological and immunohistochemical analyses of the excised lesion confirmed the diagnosis of SCL.

BACKGROUND

Spindle cell lipomas (SCLs), first described by Enzinger and Harvey¹ in the year 1975, are infrequent histological variant of lipoma, mainly presenting as well-circumscribed subcutaneous lesions of mature adipose tissue. They typically arise on the posterior neck, upper back and shoulder of elderly males between the fifth and seventh decade of life.^{1–3} They are mainly composed of mature adipocytes, bland spindle cells and collagen bundles. As it contains different components, the differential diagnosis can be very difficult, especially when they occur in other than their typical anatomical locations.

This case highlights a rare presentation of SCL in the right periorbital region of a 14-year-old boy, the MRI findings of which were suggestive of possible malignancy. The relative rarity of such a lesion in this age group and location and its misleading imaging findings and misdiagnosis on incisional biopsy were considered worthy of presentation, and hence its differential diagnosis with other tumours is discussed.

CASE PRESENTATION

The patient, a 14-year-old boy in good health, along with his parents presented with a chief problem of a slowly growing swelling around his right eye since 2 years causing the compression of the eye without inflammatory signs (figure 1). There was no history of pain, prior facial trauma or visual disturbance. The patient was moderately nourished with normal physical status and was of poor socio-economic status. The medical, surgical and family histories were unremarkable.

On examination, the swelling was smooth, irregular in shape and diffuse with no definite borders. The lesion occupied the area around the right eye extending anteriorly up to the lateral canthus of the eye, posteriorly up to the tragus of the right ear, superiorly to involve the temporal

area and inferiorly below the zygomatic arch. The overlying skin was normal; however, increased hair growth was seen around the lesion (figure 2). Eye (visual acuity, pupil reaction, ocular motility) and ears, nose and throat examinations were normal.

On palpation, the swelling was soft, non-tender and not fixed to the underlying structures or the overlying skin. There was no lymphadenopathy.

INVESTIGATIONS

Laboratory: routine blood and urine investigations were all within normal physiological range.

Radiographic investigation: orthopantomograph, postero-anterior and para nasal sinus views of face showed no abnormal findings (not shown).

CT showed well-defined, well-margined, soft tissue density lesion along the lateral wall of right orbit with the involvement of temporalis muscle. There was no evidence of bone destruction (figure 3).

MRI: coronal and axial T2-weighted (T₂W) images showed well-defined, heterogeneous area of altered signal intensity seen involving the temporalis muscle on right side along the lateral wall of the right orbit, appearing isointense to hyperintense to subcutaneous fat on T₂W₁. Surrounding

Figure 1 Photograph showing swelling around the right eye causing compression of the eye.

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Figure 2 Photograph showing swelling and increased hair growth over the skin of involved area.

fat planes were maintained. The lesion was not associated with periosteal reaction or underlying bone erosion (figure 4).

DIFFERENTIAL DIAGNOSIS

On the basis of clinical and imaging presentation, the following differential diagnoses were considered—benign neoplasm of skeletal muscle origin and possibly its malignant counterpart mainly—rhabdomyosarcoma, neural tumours mainly neurofibroma and schwannoma or a fibrous lesion.

An incisional biopsy of the lesion was conducted from the right brow region. The section showed a non-encapsulated lesion composed of interlacing bundles of elongated, spindle-shaped cells, of which some had wavy nuclei. Few small nerve fibres were present. The tumour cells were set in a fibromyxoid background. Numerous mast cells were present. The overall histopathological features were suggestive of neurofibroma.

TREATMENT

After explaining the diagnosis and surgical procedure to the patient and his parents, a written informed consent was obtained from the patient's parents (as the patient was a minor). Under general anaesthesia with orotracheal intubation, a hemi-coronal incision was given on right side and the skin flap was reflected (figure 5). Remaining superior to temporal fascia by subdermal dissection, the lesion was exposed from the superior aspect and under blunt dissection, and the entire lesion was excised taking precautions to avoid injury to superficial temporal vessels and facial nerves. The lesion was found to be unencapsulated with lipomatous component having ill-defined boundaries and was difficult to separate from the surrounding tissue. The lesion was removed in parts and involved the exploration of the entire superficial temporal space extending inferiorly up to the zygomatic arch, laterally up to the lateral wall of

the orbit and posteriorly up to the tragus of the ear. After ensuring adequate haemostasis, the lesion was closed in layers. The patient's recovery was uneventful.

OUTCOME AND FOLLOW-UP

The excisional tissue consisted of several fragments. Gross examination revealed the tumour to be rubbery with a greyish white colour and smooth surface.

Histopathological section demonstrated the admixture of spindle cells in a fibrous stroma and mature adipocytes, and there was a predominance of spindle cell component (figure 6A). The spindle cells were uniform with a single elongated nucleus and narrow bipolar cytoplasmic processes and inconspicuous nucleoli (figure 7). There was a haphazard arrangement of spindle cells, but some areas showed arrangement in short parallel bundles simulating a neural tumour (figure 6B). Few foci of concentric cells arranged in whorls were also seen (figure 6C). Occasional foci of myxoid degeneration were observed (figure 6D) and numerous mast cells (figure 7) were found to be distributed throughout the stroma. Cellular pleomorphism, nuclear hyperchromasia and mitotic figures were absent. Elongated fibroblasts elaborated a dense collagenous matrix that failed to display ropey fibres.

Immunohistochemical analysis revealed a strong and diffuse CD34 positivity in spindle cells (figure 8). The spindle cells were negative for S-100 protein (figure 9).

On the basis of histopathology and immunohistochemical findings, a final diagnosis of SCL was established. The patient made a full recovery with no neurological deficits. No recurrence was noticed after 6 months of follow-up.

DISCUSSION

SCL is a distinct entity with its characteristic clinicopathological and immunohistochemical features. It is a benign tumour which typically occurs on the posterior neck, shoulder or upper back.^{1 2} The tumour is usually well circumscribed and easily distinguished from the surrounding subcutaneous tissue. Generally, it is not encountered in children and adolescents.⁴ There is a marked male predilection especially in the sixth decade of life which may be partly explained by androgen receptor's reactivity.⁵ The findings of the present case, however, were quite contrasting with the ones above. The lesion in the present case was ill-defined with diffuse borders and occupied the right periorbital region which is not typical for SCL. Also, the patient in the present case was 14 years old which is not in accordance with the age range for SCL.

Microscopically, SCLs consist of mature adipocytes, numerous bland spindle cells, collagen fibres along with mast cells and many a times areas of myxoid degeneration. Mitosis, cellular pleomorphism and nuclear hyperchromasia are extremely rare or absent.⁶ The histopathological features of the present case fulfilled the above description. Although most SCLs are composed of a relatively equal ratio of fat and spindle cells, either of them may predominate, and the variations in the ratio of fat and spindle cells cause the wide spectrum of imaging features.⁷ In the present case, there was a predominance of spindle cell component and that could have been the possible reason for the signal intensity similar to that of skeletal muscle on T₂W images, as well as for the misdiagnosis of neurofibroma on incisional biopsy as the biopsy specimen must have contained only the spindle cell component. Another characteristic feature of SCL is the positivity of spindle cells for CD34 and negative staining with S-100 protein immunohistochemically.^{8 9} The present case showed strong and diffuse CD34 positivity of

Figure 3 Coronal CT image showing soft tissue density lesion along the lateral wall of right orbit involving temporalis muscle.

spindle cells and were negative for S-100, supporting the above finding. Mast cells are a conspicuous feature in almost all SCLs and were seen in the present case too. The significance of mast cells, however, needs to be evaluated.

Beham *et al*¹⁰ suggested the spindle cells to be of fibroblastic origin or similar to the non-lipoblastic stellate mesenchymal

cells of the primitive fat lobules which have the ability to synthesise collagen. Domanski *et al*¹¹ in their electron microscopic study showed that some of the spindle cells have some features of fibroblasts, while some contained non-membrane-bound lipid droplets, suggesting a prelipoblastic nature. However, the origin of stromal cells in SCL is still controversial.

Figure 4 Coronal and axial MRI T2-weighted images showing heterogeneous area of altered signal intensity involving the temporalis muscle.

Figure 5 Intraoperative photograph.

The histopathological differential diagnosis of SCL includes simple lipoma, fibrolipoma, pleomorphic lipoma and tumours of nerve tissue origin mainly neurofibroma and neurilemmoma. Simple lipoma do not exhibit extensive collagen bundles with spindle cells and mast cells.¹² Fibrolipomas are less cellular than SCLs and show large bundles of collagen. Pleomorphic lipomas are marked by the presence of large multinucleated floret-type giant cells with multiple radially arranged nuclei.⁶ Distinct histopathological features with positive immunostaining of spindle cells with S-100 protein helps in identifying nerve tumours and lipomatous neurofibroma.¹³

The occurrence of a spindle cell liposarcoma or a spindle cell carcinoma is excluded by the lack of mitotic activity, cellular pleomorphism and lipoblasts.¹⁴ Spindle cells' well-differentiated liposarcoma have atypical lipogenic cells showing variations in size and shape, and spindled tumour cells with slightly enlarged often hyperchromatic nuclei with mild atypia.

SCL with marked myxoid changes may be mistaken for myxoid liposarcoma as well as low-grade myxofibrosarcoma (MFS). Myxoid liposarcoma is usually deep seated and is separated from SCL by carefully estimating the vascular pattern, pleomorphism and mitotic activity together with lipoblasts.³ Immunocytochemical studies with CD34, bcl-2, 21, 24 helps in differentiating SCLs from MFS and other myxoid lesions.^{10 15}

The lesion leaves many questions unanswered:

1. What is the explanation for the predilection for the posterior neck, shoulder and upper back, although this anatomic preference has been used to support the designation of SCL as a distinct entity?
2. What is the origin of spindle cells in SCL?
3. What is the significance of mast cells in SCL?

Thus, although there is no prognostic significance in identifying this lesion as a distinct entity, a clear understanding of this lesion would help the pathologist to give definite diagnosis and differentiate it from its malignant counterparts thereby avoiding overtreatment and long-term follow-up. Nevertheless, their very rarity demands the reporting of every new case.

Figure 6 Photomicrograph showing histopathological features. (A) predominance of spindle component; (B) parallel arrangement of spindle cells; (C) whorled pattern of spindle cells and (D) myxoid area.

Figure 7 High power views showing bland and uniform short spindle cells along with mast cells in a fibrous stroma.

Figure 8 Immunohistochemistry photomicrographs showing strong and diffused positivity of spindle cells for CD34.

Figure 9 Immunohistochemistry photomicrograph showing negative staining of spindle cells for S-100 protein.

Learning points

- ▶ Biopsy is a mandatory procedure for any suspicious lesions.
- ▶ Spindle cell lipoma (SCL) is a rare benign soft tissue tumour with characteristic clinicopathological and immunohistochemical features.
- ▶ A preoperative diagnosis based on the clinical context, imaging, histopathology and immunohistochemistry is crucial for management, as it will impact the consent process, surgical approach, extent of surgery and follow-up.
- ▶ Treatment of SCL involves surgical excision with virtually no recurrences.

Competing interests None.

Patient consent Obtained.

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