CASE REPORT

Dermatofibrosarcoma protuberans of the scalp, with fibrosarcomatous areas masquerading as epidermal inclusion cyst

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SUMMARY

Lesions on the scalp are frequently encountered in clinical practice. Commonly diagnosed lesions include epidermal inclusion cysts (EICs), dermoid cysts and lipomas. Dermatofibrosarcoma protuberans (DFSP) is a low-grade malignant skin tumour occurring mostly in the extremities. However, its occurrence on the scalp is unusual, with an incidence of <5%. This lesion is rarely known to undergo fibrosarcomatous transformation. We present two such rare cases of DFSP of the scalp, with fibrosarcomatous transformation, masquerading clinically as EIC.

BACKGROUND

Painless soft tissue masses of the scalp are commonly encountered in clinical practice and include epidermal inclusion cysts (EICs), dermoid cyst, trichilemmal cysts and lipomas. Uncommonly, dermatofibrosarcoma protuberans (DFSP) can present as a scalp lesion and forms an important differential diagnosis. Furthermore, it can rarely undergo fibrosarcomatous transformation. Identifying DFSP is of utmost importance prognostically, as it differs in its management, from other commoner lesions at this site. We present two such rare cases of DFSP of the scalp, with fibrosarcomatous transformation, masquerading clinically as an EIC.

CASE PRESENTATION

Case I: A 23-year-old man presented to surgical outpatient department (OPD) in 2006 with an exophytic, painless mass measuring 4×2 cm in the right occipital region for the past 3 years. On examination, the lesion was firm in consistency and mobile. The mass was recurrent with history of excision 3 years prior at the same site, records of which were not available. A provisional diagnosis of EIC was made and lumpectomy performed and sent for histopathological examination (HPE). Microscopic examination showed the presence of proliferation of spindle-shaped cells arranged in a storiform pattern reaching deep to the level of subcutaneous tissue, and entrapping it. No atypia/mitosis/necrosis was noted. The lesion was involving the margins. Immunohistochemistry (IHC) showed positivity for vimentin and CD34. On the basis of these findings, a diagnosis of DFSP was made. Since the margins were involved, the patient was advised close follow-up. Ten years later, in 2016, he presented again, with another recurrence, this time as a multinodular, exophytic growth at

the same site. CT scan revealed no intracranial extension. Wide local excision (WLE) was performed and the specimen sent for HPE. On gross examination, a skin covered multinodular lesion measuring 9×5 cm was seen. On cut section, the lesion was firm and greyish-white ([figure 1](#)). Microscopic examination revealed a cellular spindle cell lesion arranged in fascicles with a herring bone pattern, having mild-to-moderate atypia and 1–2 mitosis/high power field (HPF) ([figure 2A](#)); however, no necrosis was noted. The adjacent areas showed features of DFSP showing spindle cells arranged in storiform pattern with neither atypia nor mitosis. IHC in the cellular areas showed diffuse positivity with vimentin ([figure 2B](#)) but CD34 expression was lost ([figure 2C](#)). CD34 expression was, however, retained in the DFSP-like areas ([figure 2D](#)). On the basis of high cellularity, mitosis and loss of CD34 expression, a diagnosis of fibrosarcomatous change in DFSP was offered. All the margins were free of tumour.

Case II: A 28-year-old woman presented to surgical OPD with a painless, polypoidal lesion on the left scalp for the past 2 months. On examination, it measured 2×2 cm and was firm and non-tender ([figure 3](#)). A provisional clinical diagnosis of EIC was made; the swelling was excised and sent for HPE. Microscopy revealed the presence of a cellular spindle cell lesion in the dermis, with cells arranged in a storiform pattern. The centre of the lesion showed high cellularity with increased mitosis of 1–2/HPF ([figure 4A](#)), with focal areas of necrosis. The periphery of the lesion showed spindle cells arranged in a storiform pattern with neither atypia nor mitosis ([figure 4B](#)). IHC revealed diffuse vimentin positivity ([figure 4B inset](#)) with the presence of CD34 expression only at the periphery ([figure 4C](#)), however, CD34 expression was lost in the centre where cellular, mitotic rich areas were present ([figure 4D](#)). All the margins were free of tumour and the lesion was just short of the deep resected plane. Thus a final diagnosis of DFSP with focal fibrosarcomatous transformation was made.

CT scan was performed only in our first case, which showed the lesion limited to the soft tissue, with no underlying bony erosion and no metastasis. The second patient was unwilling to undergo further investigations.

DIFFERENTIAL DIAGNOSIS

DFSP is a low-grade malignant tumour comprising <0.1% of all malignancies.^{1 2} The lesion is more

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Figure 1 Gross skin covered multinodular lesion. Cut section is firm and greyish-white.

common in the second and third decades,³ and occurs mainly over the trunk and proximal extremities.⁴ The scalp is a rare site of involvement, with <5% incidence.^{5 6} These tumours are usually superficial in this location and are variably sized, nodular and exophytic in appearance.⁷ Therefore, they can be easily mistaken clinically for commonly encountered lesions at this site, such as EICs, dermoid cysts, trichilemmal cysts and lipomas.

EICs and dermoid cysts, though actually cystic lesions, often appear firmer in consistency due to the semisolid nature of their contents, which further adds to the confusion.⁸ Trichilemmal cysts, though uncommon on the scalp, often present as variably sized, exophytic masses with presentation very similar to DFSP.⁹ Lipomas, especially the spindle cell variant,¹⁰ and liposarcomas,¹¹ are other rare lesions that can present on the scalp and appear grossly similar.

Fine-needle aspiration cytology (FNAC) can aid in preoperative diagnosis and is recommended in all cases. Unfortunately, in both of these cases, FNAC was not carried out.

All these lesions can be easily diagnosed on histopathology on the basis of granular layer, with its absence diagnostic of trichilemmal cysts and presence indicative of either EICs or dermoid cysts, which can be further differentiated depending on whether

Figure 3 Gross firm greyish-white lesion excised as an epidermal inclusion cyst.

the adnexal structures are seen in the cyst wall or not. Spindle cell lipomas show the presence of spindle cells surrounding fibrous tissue without atypia. The presence of atypia with lipoblast against a fatty background indicates a possibility of liposarcoma. DFSP, on the other hand, shows proliferation of spindle cells in a storiform pattern engulfing the subcutaneous tissue with no mitosis/atypia/necrosis.

Although histopathology is required for establishing diagnosis and DFSP is rare on the scalp, it would be prudent for clinicians to keep it in mind as a differential diagnosis, especially in cases of recurrent lesions. All the lesions occurring frequently at this site are benign and require simple excision, whereas DFSP, having a higher probability of recurrence and potential to turn malignant, requires WLE. Increased cellularity with a herring bone pattern, mild-to-moderate atypia and mitosis >1–2/HPF should alert the pathologist to the possibility of fibrosarcomatous transformation in DFSP, characterised by loss of CD34 expression on IHC.¹²

Figure 2 Cellular spindle cell lesion with cells arranged in herring bone pattern with mild-to-moderate atypia (H&E, $\times 400$). Inset showing a mitotic figure (H&E, $\times 600$) (A). Diffuse positivity with vimentin seen in the cellular areas ($\times 400$) (B). Loss of CD34 expression in cellular areas indicating fibrosarcomatous transformation. Endothelial cells of blood vessel acting as positive control ($\times 400$) (C). CD34 expression seen in dermatofibrosarcoma protuberans-like areas ($\times 400$) (D).

Figure 4 Skin lined biopsy showing cellular spindle cell lesion in the dermis arranged in a storiform pattern (H&E, $\times 400$). Inset: showing mitotic figure in the lesion (H&E, $\times 600$) (A). Spindle cells arranged in storiform pattern with neither atypia nor mitosis seen at the periphery in dermatofibrosarcoma protuberans-like areas (H&E, $\times 400$). Inset: showing diffuse vimentin positivity in these areas ($\times 400$) (B). Positive CD34 expression at the periphery in dermatofibrosarcoma protuberans-like areas ($\times 400$) (C). Loss of CD34 expression in cellular, mitotic rich areas in the centre indicating fibrosarcomatous transformation ($\times 400$) (D).

The differential diagnosis based on histology includes lesions showing a storiform pattern and usually falling under the category, fibrohistiocytic tumours. Atypical benign fibrous histiocytoma (BFH) acts as a close differential, as seen in our case, as it also shows presence of spindle-shaped cells arranged in a storiform pattern with occasional mitosis and necrosis. However, marked atypia and presence of bizarre cells characteristic of atypical BFH are absent in conventional DFSP; nuclear atypia is higher and mitotic figures more numerous in the fibrosarcomatous component.

OUTCOME AND FOLLOW-UP

The patient in our first case, 6 months post-WLE surgery, is currently doing well with no recurrence of lesion. The patient in

our second case was unwilling to undergo repeat surgery for marginal clearance and was later lost to follow-up.

DISCUSSION

Currently, <200 cases of DFSP turning malignant at various sites have been reported in the literature, with only a handful of cases involving the scalp.^{4 13 14} DFSP with malignant transformation on the scalp has been known to metastasise to distant sites such as the lungs.¹³ Therefore, these patients should be kept on close follow-up. In addition, cases of scalp DFSP are rarely reported to cause intracranial extension.¹⁴ Therefore, radiological investigation must be performed in all cases to find out the extent of involvement.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Dermatofibrosarcoma protuberans (DFSP) of the scalp is rare, with <5% incidence.⁵
- ▶ A <200 cases of DFSP turning malignant at various sites have been reported in the literature, with only a handful of cases involving the scalp.^{4 13 14}
- ▶ Clinicians must keep DFSP as a differential while evaluating scalp lesions irrespective of their size, particularly in recurrent cases.
- ▶ Fine-needle aspiration cytology can be used as a screening test for preoperative diagnosis and wide local excision should be performed in suspicious cases. Radiological assessment for extent of involvement by the lesion must be determined.
- ▶ Malignant transformation can occur even in small-sized lesions therefore pathologists should sample all cases of DFSP extensively and look for areas of high cellularity, mitosis, atypia and necrosis, to rule out such a possibility. In addition, loss of CD34 expression can aid in diagnosis.

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