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Case Report

Deep benign fibrous histiocytoma of the retropharyngeal region - A rare case report

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Abstract

Deep benign fibrous histiocytoma (DBFH) is a rare, non-cancerous soft tissue tumor that typically arises in deep dermal or sub-cutaneous tissues. It is characterized by combination of fibroblastic and histiocytic cells. DBFH predominantly affects adults, with a preference for the limbs and trunk. The tumor often presents as a slow-growing, painless mass that may remain undiagnosed for a long period of time due to its indolent nature. DBFH should be differentiated from other soft tissue tumors, including malignant variants. This case report aims to present a rare instance of DBFH in a 50-year-old female with unusual site of presentation along with providing insights into its clinical presentation, diagnostic challenges, and treatment options.

DBFH is a rare, benign tumor with a slow progression and a low recurrence rate if properly excised. It poses diagnostic challenges due to its similarity with malignant tumors. Surgical excision remains the primary treatment, and close follow-up is essential to monitor for recurrence. Although the tumor follows an indolent course, accurate diagnosis and differentiation from malignant soft tissue tumors are crucial.

Keywords: Mesenchymal, Dermatofibrosarcoma protuberans, Soft tissue, Immununohistochemistry, Benign.

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1. Introduction

Deep benign fibrous histiocytoma (DBFH), also known as deep fibrous histiocytoma (DFH), is a rare, benign soft tissue tumor that can occur in subcutaneous or deep soft tissues, and is composed of fibroblasts and histiocytes. While most commonly found in subcutaneous or deep soft tissues, it can also occur in the extremities and head and neck.[1]

These tumours are well circumscribed with mixed fascicular and storiform growth pattern, consisting of monomorphic histiocytoid or spindled cells interspersed with branching vessels.

DBFHs are typically slow-growing and painless. Males are slightly more affected than females.

Although these are benign tumours, local recurrence occurs in upto 20% of cases.[2,3] Distant metastasis is rare.

Diagnostic criteria according to the WHO classification of soft tissue and bone tumors (5th edition):[4]

1. Well-defined lesion in subcutaneous or deep/visceral tissue
2. Mixed fascicular and whorled/storiform growth patterns
3. Monomorphic spindle or histiocytoid cells
4. Branching hemangiopericytoma-like vascular pattern

The treatment of deep fibrous histiocytomas usually is surgical excision. We hereby present a rare case of DBFH in the prevertebral location in a middle aged female.

2. Case Presentation

A 50-year-old woman presented to the outpatient clinic with a two month history of painless, progressively enlarging neck swelling. She had no history of trauma or any related medical /surgical issues. On examination, a non-tender, mobile, firm mass was noted in neck. The patient had no difficulty in swallowing. Ultrasound revealed a large, heterogeneously hypoechoic lesion measuring 16 cm in diameter. It was located posterior to trachea & esophagus, and extending bilaterally in the neck, superiorly upto the mandible and

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inferiorly just above the sternum. The mass caused anterior displacement of trachea, esophagus& carotid vessels. MRI showed homogenously enhancing solid soft tissue mass lesion in the pre-vertebral space of neck, extending inferiorly into posterior superior mediastinum, exerting a mass effect on surrounding structures.

With informed consent, the mass was completely excised via wide local excision. The excised specimen measured 16x15x6 cm and was partially encapsulated. It was soft to firm with grey white to yellowish cut surface and showed areas of necrosis& hemorrhage.

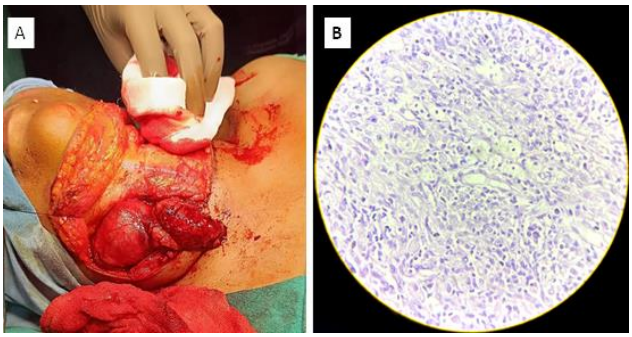

Figure 1: A: Intra-operative image of the neck mass; **B:** Microscopic image showing bland spindle cells arranged haphazardly along with scattered histiocytes (H&Ex400).

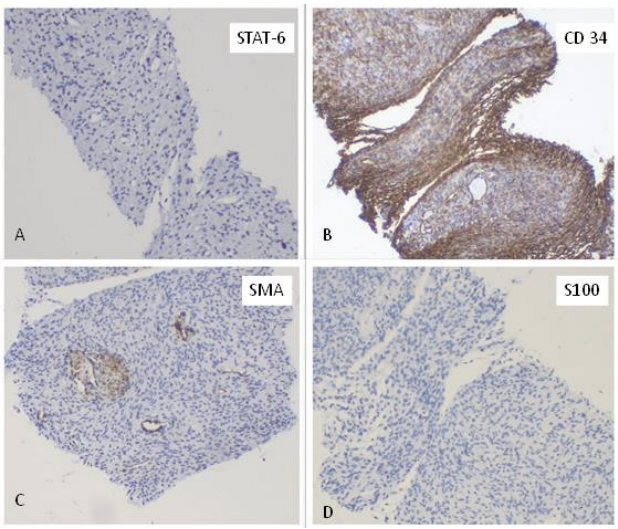

Figure 2: Immunohistochemistry shows tumour cells positive for CD34 (B) and negative for other markers.

Histology revealed haphazardly arranged fascicles of spindle cells with moderate anisonucleosis & variably prominent nucleoli. Clusters of foamy macrophages & lymphocytes were observed, along with focal coagulative necrosis. Mitotic activity was low (0-1/10 hpf). Immunohistochemistry studies showed tumor cells were positive for CD34, while negative for SS18-SSX, SMA, Desmin, STAT6, ERG, CD117, DOG1 and MUC4. The Ki67 proliferation index was low and was 5%. A final diagnosis of DBFH was made after ruling out other differentials.(**Figure 1**, **Figure 2**)

3. Discussion

DBFH is a tumor of unknown origin composed of fibroblasts and histiocytes. It was first described by Dahlin in 1978 and accounts for only 1-2% of all fibrous histiocytomas.[2,3] These tumors can be anatomically classified into cutaneous and deep forms. Cutaneous form has many variants such as cellular, atypical and others. Unlike the cutaneous counterpart, DBFH tends to be well circumscribed, occasionally encapsulated, and typically exhibits uniform, benign spindle cells in a storiform pattern. These cells have tapered oval nuclei with fine chromatin, one to two small nucleoli and rare mitosis (less than 5/10 high power fields). However, some cases might show mixed cellularity composed of foamcells, siderophages, mast cells, lymphocytes and osteoclast-like giant cells, similar to the cutaneous BFH. Tumor necrosis and cellular pleomorphism are extremely rare.[5] In our case, necrosis was present, but it was coagulative type of necrosis which may be due to large tumour size. Mitotic activity was scanty in our case along with low cellularity, differentiating it from other malignant tumours. Branched, staghorn, dilated blood vessels (hemangiopericytomatous-like) are occasionally seen along with central cystic degeneration, and stromal hyalinization, features that do not occur in the cutaneous counterpart.[5-7] Immunophenotypically, cutaneous BFH and DBFH share positive expression of Factor XIIIa, however the former is typically negative for CD34 and the later exhibits CD34 positivity in 40% cases.

Diagnosis of DBFH relies on histopathological evaluation of the excised mass, as its clinical, radiological, and histological features can closely resemble those of other benign and malignant tumors. Differential diagnoses include schwannoma, solitary fibrous tumor (SFT), dermatofibrosarcoma protuberans (DFSP), leiomyosarcoma, and malignant peripheral nerve sheath tumor (MPNST).

Schwannomas can be readily distinguished from DBFH by their thick, hyalinized blood vessels, peripheral lymphocytic cuffing, hypercellular areas with nuclear palisading (Verocay bodies), and strong diffuse expression of S100 protein, features not seen in DBFH. Similarly, MPNSTs exhibit hypercellularity, nuclear atypia, increased mitotic activity, and focal S100 expression, all of which are absent in DBFH.

DFSP was one of the key differentials considered in our case. However, DFSP is a dermal-based, infiltrative, hypercellular neoplasm composed of monomorphic malignant spindle cells that infiltrate surrounding fat and collagen in a honeycomb pattern, and it demonstrates diffuse CD34 positivity. In contrast, the lesion in our case was located in the prevertebral region, with no cutaneous involvement, low cellularity, absence of a storiform pattern,

and scant mitotic activity. Furthermore, while up to 40% of DBFH cases express CD34, our case lacked the diffuse positivity typical of DFSP.[8]

Solitary fibrous tumor (SFT) was also considered, but it was excluded due to the absence of STAT6 expression. Leiomyosarcoma was ruled out based on negative immunohistochemical staining for desmin, SMA, MSA, and h-caldesmon. Additionally, the absence of nuclear atypia and mitotic activity supported the benign nature of the tumor.

The recommended treatment for DBFH is wide local excision, followed by close monitoring, as local recurrence can occur in up to 20% of cases if excision is incomplete.[2] Although DBFH is considered benign, Gleason and Fletcher have reported occasional, unpredictable metastatic potential.[3]

4. Conclusion

In conclusion, based on our findings and review of the literature, DBFH generally follows an indolent course with a low risk of recurrence. However, due to its clinical and histological resemblance to malignant soft tissue tumors, accurate diagnosis and thorough histopathological evaluation are essential to guide appropriate management.

5. Source of Funding

None.

6. Conflict of Interest

None.

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