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# DEEP BENIGN FIBROUS HISTIOCYTOMA OF THIGH – A CASE REPORT WITH REVIEW OF LITERATURE

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**ABSTRACT**

**Background:**Benign fibrous histiocytoma is a mesenchymal tumor that is believed to originate from fibroblasts and histiocytes. It is classified anatomically into Cutaneous and Deep forms. Deep benign fibrous histiocytoma (DBFH) is rare tumor accounting from 1% to 2% of all benign fibrous histiocytoma. It is well circumscribed, occasionally capsulated and typically shows uniform, benign spindle cells arranged in 'storiform pattern'. It can develop at any age, commonly affects middle aged males.

**Case details:**A 13-year-old female child presented with painless swelling in left thigh since 02 years. On local examination a single swelling of approximately 13 x 10 cm in size was palpated over lateral aspect of left thigh which was elastic, movable and non-tender on palpation.

**Investigations and diagnosis:** Ultrasound showed well circumscribed heterogeneously hypoechoic lesion in lateral aspect of left thigh.

Trucut biopsy was performed and on HPE probable diagnosis of Low grade malignant peripheral nerve sheath tumor was made. Later excision biopsy was done, histopathology and IHC studies confirmed the diagnosis of Deep benign fibrous histiocytoma.

**Conclusion**:Deep benign fibrous histiocytoma is a very rare entity which cannot be diagnosed clinically. DBFH follows an indolent course and has a low risk of recurrence. Although challenging to diagnose clinically and histopathologically, it needs to be accurately distinguish from malignant soft tissue tumor.

## INTRODUCTION

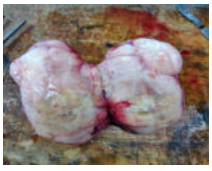
Benign fibrous histiocytoma (BFH) is a benign mesenchymal tumor believed to be developed from fibroblasts and histiocytes. Anatomically, BFH is divided into two types: cutaneous and deep. Deep benign fibrous histiocytoma (DBFH) is a rare benign fibrous histiocytoma that accounts for about 1% to 2% of all BFHs [1]. Although it can afflict anyone at any age, DBFH is more common among middle-aged men. DBFH is characterized by a slow-growing, painless mass that affects the extremities, head, and neck [3]. We report a case of approximately 12 cm DBFH found in thigh of young patient describing its clinical features, radiological evaluation, histopathologic, and immunophenotypic characteristics.

### Case report

A 13-year-old female child presented with painless swelling in left thigh since 02 years. On local examination a single swelling of approximately 13 x 10 cm in size was palpated over lateral aspect of left thigh, which was elastic, movable and non-tender on palpation. Ultrasound revealed a well-defined heterogeneously hypoechoic lesion in antero-lateral aspect of left thigh. Tru-cut biopsy was performed and on HPE probable diagnosis of Low grade malignant peripheral nerve sheath tumor was made. Later excision biopsy of the mass done followed by immunohistochemistry. Final diagnosis of deep benign fibrous histiocytoma was made.

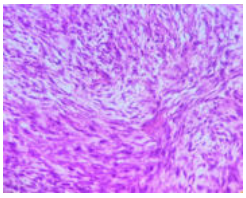
### Gross examination

the tumor showed a encapsulated soft tissue mass of approximately size 12x9x6 cms, attached with fibro-fatty tissue. On cut, multiple nodules of variable sizes seen which were homogeneous white in color. We also received a fibrofatty tissue of 5x2x0.5 cm yellowish white in color. single lymph node identified and dissected measuring 2x1x0.1 cm.



Encapsulated soft tissue mass of approximately size 12 x 09 x 06 cms, attached with fibro-fatty tissue.

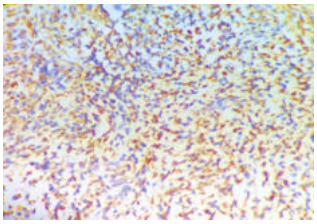
### Microscopically



Multiple sections studied from excised mass revealed uniform cellularity of spindle cells predominantly arranged in 'Storiform' pattern and few arranged in 'Herring bone' pattern. Spindle cells had ill-defined eosinophilic cytoplasm, elongated plump nuclei with tapered ends. Few dilated blood vessels and scattered lymphocytes seen in the stroma. Occasional areas of hemorrhage and necrosis seen. No mitosis seen. Sections from the lymph nodes shows intact capsule and normal nodal architecture. There are numerous

lymphoid follicles seen of variable sizes with prominent germinal centre. Dilated sinusoids which are filled with histiocytes. Numerous dilated blood vessels seen in medulla.

**Immunohistochemistry** Reveals the following results.
Desmin – negative
Vimentin – positive
S-100 – negative



**Literature review**
We searched database (PubMed) for case series, reports and review studies related to deep benign fibrous histiocytoma of the soft tissue. We chose articles from 2010 to 2021.

**Abrar J filfilan** et al shows a case of a 31-year-old man who lived with a painless mass in his right thigh for 15 years. After its examination by preoperative ultrasound and magnetic resonance imaging, the tumor was completely excised. Further histologic evaluation revealed neoplastic growth composed of proliferating spindle cells forming short fascicles and a storiform pattern. The patient was diagnosed with DBFH on the basis of its intraoperative, radiologic, and histopathologic features

**Angelica Puopolo** et al presents the case of a 43-year-old man smoker who was taken to a fast access lung clinic with hemoptysis, chest discomfort, and axillary lymphadenopathy, a clinical picture that triggered suspicions of an underlying cancer. A right hilar mass, multiple parenchymal cysts, and a massive mediastinal mass were discovered on a computed tomography (CT) pulmonary angiography. The mediastinal mass was diagnosed as a low-grade spindle cell tumour after a CT-guided biopsy. The lump was surgically resected due to its massive size, and it was determined to be a deep benign fibrous histiocytoma. The importance of this study is to highlight a clinical presentation that appears to be malignancy but is actually an uncommon variety of a benign tumour.

**Gleason et al**
published the largest single series in the English literature in 2008, they reported the clinical and pathological features of 69 cases of DBFH (Table 1). 28 patients from this series has a preoperative periods ranging from 3 weeks to 10 years and the most common anatomical site was the lower extremity (33%) with lower legs being the commonest subsite. Other documented case reports of DBFH in the extremity along with rare anatomical locations in visceral organs, showed sizes ranging from 0.9 to 23 cm, and majority developed over a preoperative period ranging from 2 months to 2 years, with wide age range from 19 to 63 years (Table 1).

**Akublut et al**. reported a case of 3 × 2.5 cm DBFH in the shoulder developed over 15 years. As seen with our patient, the DBFH is a slow growing lesion, with only 12 cm of growth over a 5-year period [7]

**Vijayasaradhi Annam et al** A 25-year-old woman with a history of dry cough, intermittent fever, and chest pain over the past year was reported. A CT scan of the posterior mediastinum on the right side revealed a massive, well-defined, heterogeneous solid mass lesion. A diagnosis of benign posterior mediastinal mass, presumably neurogenic tumour with emphysematous bullae, was made based on these findings. The results of fine needle aspiration cytology conducted under CT guidance were inconclusive. Following that, a right-sided thoracotomy was conducted, revealing a well-encapsulated soft tissue mass that was immediately distinguishable from the paraspinal soft tissues and unrelated to the lung, pleura, oesophagus, and other mediastinal structures. The patient's postoperative course was uneventful. On histology, a final diagnosis of benign fibrous histiocytoma was made.

| Serial no | Study | Age | Sex | Year | Anatomical site | size | treatment |
|---|---|---|---|---|---|---|---|
| 1 | Abrar J filfilan et al | 31 | M | 2019 | Thigh | 8.5x10 | Excision |
| 2 | Angelica Puopolo et al | 43 | M | 2017 | Ant mediastinum | 0.9cms | surgery |
| 3 | Eun jo et | 36 | F | 2015 | Buccal mucosa | 2.3x1.3x1.3 cms | excision |
| 4 | Akublut et al. | | | | | | |
| 5 | Vijayasaradhi Annam et al | 25 | F | 2010 | Posterior mediastinum | 12x9.6x7.8cms | Excision |
| 6 | Gleason et al | Median age 37 years | M >F | 2008 | Extremities | - | surgery |

**DISCUSSION**
DBFH is a tumor made up of fibroblasts and histiocytes with an unclear cause. Anatomically, these tumors can be divided into two types: cutaneous and deep. Deep form is uncommon, accounting for just 1 to 2% of all BFHs [6,9]. Cutaneous forms come in a variety of forms, including cellular, atypical, and others [10]. Cutaneous cellular fibrous histiocytoma is a hypercellular, uncapsulated tumor that can spread to the superficial subcutaneous tissue. This tumor is made up of short and long fascicles of homogenous spindle cells with no storiform pattern, hypercellularity in the core and hypocellularity in the periphery, and collagen entrapment is a key diagnostic feature [10]. DBFH tends to be well circumscribed, capsulated on occasion, typically shows homogenous, benign spindle cells organized in a storiform or herring bone pattern. These cells feature tapering oval nuclei with thin chromatin, one to two tiny nucleoli, and only 5/10 high power fields of mitosis [10]. Some cases, like cutaneous BFH, may display a mix of cellularity that includes foam cells, siderophages, mast cells, lymphocytes, and osteoclast-like giant cells. On immunohistochemistry, both cutaneous BFH and DBFH express Factor XIIIa, but the former is always negative for CD34, whereas DBFH may express it. Because it shares clinical, radiological, and histological features with other benign and malignant tumors such as schwannoma, solitary fibrous tumour (SFT), dermatofibrosarcoma protuberans (DFSP), leiomyosarcoma, and malignant peripheral nerve sheath tumour, DBFH is only confirmed through histopathologic evaluation of the excised mass (MPNST). Schwannoma may clearly be distinguished from DBFH because it has thick hyalinized blood vessels, capsular lymphocytic cuffing, hypercellular regions with nuclear palisading, and robust diffuse S100 expression. Hypercellularity, nuclear atypia, increased mitotic activity, and localized expression of S100 are all seen in MPNST. DFSP is a hypercellular dermal tumor formed of monomorphic malignant spindle cells that entrap the surrounding fat and collagen in a honeycomb pattern and are diffusely CD34 positive. Although SFT is the most common differential diagnosis for DBFH, our case was distinguished from that of STF by the absence of round to oval spindle cells arranged in a disorganised patternless pattern with alternating hypercellular and hypocellular areas, dilated blood vessels with thick hyalinized walls, and negative CD34 immunohistochemical expression. Although immuno histochemistry is not important in the diagnosis of DBFH,

CD34 and SMA are positive in around 40% of cases, making it even more difficult to distinguish DBFH from SFT [12,13]. DBFH is best treated with a large local excision and diligent monitoring, as it can return locally in up to 20% of patients if not thoroughly excised [9]. In conclusion, DBFH has an indolent course and a minimal chance of recurrence, based on our case and the literature. It must be recognized from malignant soft tissue tumours, despite being difficult to diagnose clinically and histopathologically.

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