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Plexiform neurofibroma: An Arduous entity

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ABSTRACT

Plexiform neurofibroma being the uncommon variant of neurofibroma generally involves head and neck region. It arises from the proliferation of all neural elements. Presence of this tumor at lower extremity is highly uncommon and involves lots of complications. In this study we have taken highly vascular nature of the tumor in consideration and done prior histopathological confirmation along with immunohistochemistry marker to confirm the diagnosis. Proper prior examination involved radiological screening and colored Doppler investigations. Previous failed surgery of patient marked into the importance of present concern regarding the surgery. Uncommonly, a plexiform neurofibroma may change into a cancer, called a malignant peripheral nerve sheath tumor (MPNST). There are no reliable tests to screen for an MPNST. Thus surgery was performed and specimen has been sent to histopathology. Diagnosis was confirmed with the help of IHC and patient was discharged after recovery from surgery.

Keywords: Plexiform neurofibroma, Nerve sheath tumor, neurofibromatosis type I, Schwann cell tumor, CD34 marker

1. INTRODUCTION

Plexiform neurofibroma is an uncommon variant of neurofibroma (Weiss et al., 2007). It is the benign peripheral nerve sheath tumor and uncommonly seen in neurofibromatosis type. These lesions are mostly seen in young adults, without any gender predilection. Lesions are composed of Schwann cells, perineurial cells, fibroblasts, and mast cells (Lee et al., 2012a; Varma et al. 2020). Two to five % of these tumors also have malignant potential (Savva et al., 2010).

Neurofibromas are most commonly observed on the head and trunk. While the vertebral column, mandible, oral mucosa, male breast, subungual space, retroperitoneum, lips, and scrotal sac are the uncommon sites (Lee et al., 2012b). Isolated plexiform neurofibroma of the thigh is an extremely rare finding. To our best knowledge, only a few cases are reported (Tahririan et al., 2014b; Liu et al. 2019). With this, we report an unusual case of plexiform neurofibroma of the thigh. The diagnosis is based on clinical presentation, imaging, and histopathological findings.

2. CASE REPORT

Clinical presentation

A 45 years male presented with painless swelling over the right thigh for the last ten years. The swelling was insidious in onset; initially swelling was about 4x6 cm in size towards the outer aspect of midthigh, which progressed to the current size of 40x30 cm.

Previous treatment

There was a history of excisional surgery five years ago, but couldn't be accomplished because of uncontrolled bleeding

Radiological examination

Contrast-enhanced CT scan with angiography

There is soft tissue attenuation of a complex solid cystic lesion in the deep intramuscular plane measuring approximately 19 x 14 x 26 cm over the right tight. The lesion as shown in radiological images figure 1 and figure 2 is showing moderate enhancement of the solid component along the periphery of the lesion with the necrotic component within it. The lesion is predominantly fed by the branches of the profunda femoris artery. The mass is also displacing and compressing profundal femoris medially.

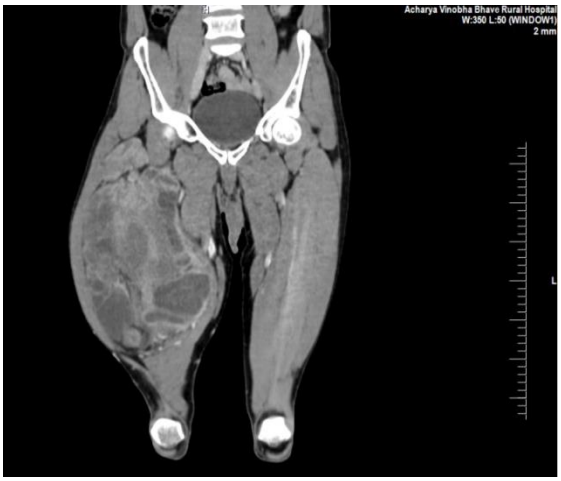

Figure 1 CT angiography of lower limb

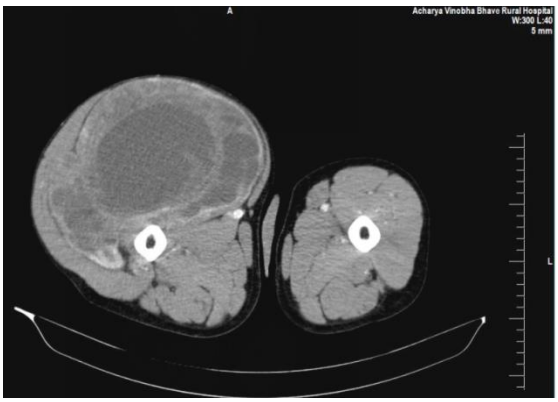

Figure 2 CT angiography of lower limb

Current Treatment

After routine blood investigations and CT scan of the tumor, surgical resection of the tumor was done, total surgical resection of the lump is shown in figure 2. The proper care of excessive bleeding and adjacent structures were taken with fluid and blood transfusion during the surgery.

Histopathology Report

Gross findings

We received a single, irregular, multi-loculated tissue piece measuring 21 x 20 x 8 cm. On cut section of tissue-piece showed a single cystic cavity with homogenous reddish material, cystic cavity measuring 13 x 12 cm.

Microscopic Findings

As shown in figure 3 section shows epidermal and subcutaneous tissue, profusely expanded in the background of hypocellular myxoid material. The occasional Schwann cells, mast cells, and fibroblasts were also seen. These histopathological features were suggestive of plexiform neurofibroma (figure 4).

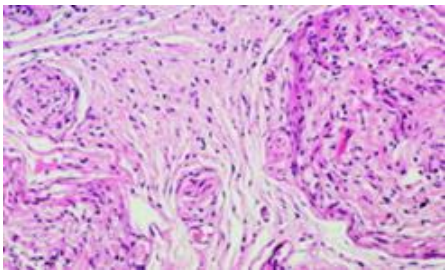

Figure 3 Microscopic findings under 100x magnification

Immunohistochemistry

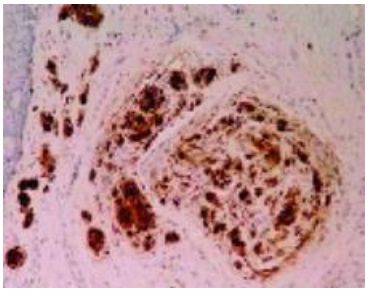

Figure 4 CD34 marker also showed intense positivity.

3. DISCUSSION

Out of all the subtypes, plexiform neurofibroma occurs in approximately 26.7% of cases (Huson et al., 1988). Presenting as the excessive skin pigmentation, hyperkeratosis and abnormal hair growth, these are some of the common factors in inborn plexiform neurofibroma (Gutmann et al., 1997). Being one of the rarest subtypes in neurofibroma and present at an abnormal site, proper diagnosis is necessary for the management and control of disease for any further deformity.

Plexiform neurofibroma in some cases shows highly aggressive nature while remain silent in others (Woodruff 1999; Re et al., 2003). These neurofibromas generally do not present in lower extremities. Plexiform neurofibroma present in calf region was then diagnosed on histopathology in this case report. Also to determine the accuracy of test application of CD 34 marker is being used. Its result came intensely positive. Plexiform neurofibromais found in most of the cases should be diagnosed and treated early had better prognosis (Friedrich et al., 2005; Needle et al., 1997). Plexiform neurofibroma by presentation can mimic hemangioma and other soft tissue tumors. Histological examination and immunohistochemistry optimise the diagnosis.

4. CONCLUSION

Plexiform neurofibromatosis is one of the rarest subtypes of neurofibroma. The distinct clinical, histopathological, and radiological findings suggest the diagnosis of plexiform neurofibroma. Figure 4 shows the intense positivity for immunohistochemistry with CD34 marker confirms the diagnosis.

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Conflict of Interest

The authors have no conflict of interest.

Informed Consent

Written and informed consent was taken from the patient, and additional informed consent was informed from the patient for whom identifying information is included in this manuscript.

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Data and materials availability

All data associated with this study are present in the paper.

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