



A LARGE SCALP MYXOID NEUROFIBROMA AN UNUSUAL PRESENTATION IN A MIDDLE- AGED PATIENT: A RARE CASE REPORT

Dr. Subhabrata Das	Professor, Department of General Surgery, R G KAR Medical College and Hospital, 1 Khudiram Bose Sarani, Kolkata 700004, India.
Dr. Mala Mistri	Associate Professor, Department of General Surgery, R G KAR Medical College and Hospital, Khudiram Bose Sarani, Kolkata 700004, India.
Dr.Sukanta Sikdar*	Faculty, Department of General Surgery, R G KAR Medical College and Hospital, 1 Khudiram Bose Sarani, Kolkata 700004, India.*Corresponding Author

ABSTRACT The transformed cells in a neoplasm, whether benign or malignant, often resemble each other, as though all had been derived from a single progenitor, consistent with the monoclonal origin of the tumor. Myxoid neurofibroma (MN) is a benign tumor of perineural origin, which is demonstrated by positive immunohistochemical staining for S100 protein. The most common locations are the face, shoulder, anus, periungual, and feet. *To our knowledge, this is the first report of an MN in the scalp, which is a very rare location that has been reported earlier. The differential diagnosis of the tumor at this location MN should be kept in mind.* This 56 years old male who presented with a large swelling in the scalp (occipital region) which extended to the nape of neck for last 3 years which is gradually increasing in size along with heaviness, intermittent severe pain in the head. Clinically (25x20) cm size swelling in the occipital area and extending to the nape of the neck. The swelling is nontender. It is ovoid in shape. Soft cystic in consistency, the surface is smooth, margins are well defined, the mobility is absent. Fluctuation test is negative but the swelling is brilliantly transilluminant.

CONCLUSION: *We report this case because of the rarity of both the tumor and its scalp location and also a giant size and to provide a review of the literature.* This case study illustrates that any slowly progressing swelling in an unusual location should have been properly investigated and complete surgical excision is the preferred choice of treatment for future recurrence. The MN should be included in the differential diagnosis of tumors at this location.

KEYWORDS : MN -Myxoid Neurofibroma, SOL – Space Occupying Lesion, HPE –Histopathlogical Examination, Excision Of Cyst, MRI – Magnetic Resonance Imaging

1. INTRODUCTION:

In some unusual instances, however, the tumor cells undergo divergent differentiation, creating so-called mixed tumors. Solitary neurofibroma can present as one of the following variants: cutaneous lipomatous, collagenous, epithelioid, granular, pigmented, dendritic cell, and myxoid neurofibromas [1]. We present this case of a myxoid neurofibroma (MN) of the scalp, which is a rare location of this tumor with an unpredictable presentation.

2. CASE PRESENTATION: The patient is 56 years old male with a low social background with diabetes and COPD who presented with a large swelling in the scalp (occipital region) for the last 3 years which is gradually increasing in size along with heaviness, intermittent severe pain in the head but no history of tingling sensation, nausea, vomiting or vertigo.

The patient had a past history of cataract surgery but presently without any problem. Clinically patient has (25x20) cm swelling in the occipital region and extending to the nape of the neck. It is ovoid. Soft cystic in consistency, the surface is smooth, margins are well defined, The mobility is Fluctuation test is negative but the swelling is brilliantly transilluminant.

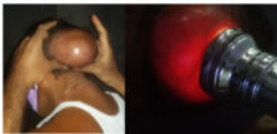


Figure 1. Clinical examination - Soft cystic swelling, absent mobility, and brilliantly transilluminant.

CT scan of brain showing large well defined soft tissue density SOL (average +17 HU) noted in the left occipital region of the scalp. No obvious calcification was noted within the SOL.

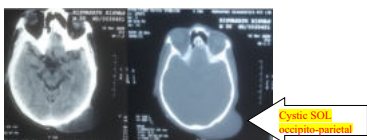


Figure 2. CT scan of brain (plain) -Soft tissue SOL without any bony defect in left of occipito-parietal area.

MRI of brain showing a large, well defined thick-walled cystic SOL is seen involving the subcutaneous tissues of the scalp and extending into the nape of the neck on the left side. No solid component noted within the SOL, No loculi and internal septation seen. The SOL measures 74.4 mm (AP) x 117.3 mm (ML) x 106 mm (SI). It is seen to be confined within the subcutaneous tissues of the neck and is seen to cause mild indentation over the adjacent muscles. No infiltration between the intramuscular planes is seen. No significant enlargement of cervical lymph nodes seen.

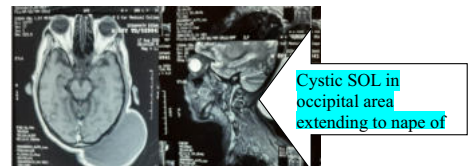


Figure3. MRI of brain (plain) -Cystic SOL in scalp at occipital region and extending to nape of neck.

As the patient is being suffered from COPD, we did PFT and optimize the patient with nebulization with levolin and budesocort inhaler and incentive spirometry and control of blood sugar with OHA before the operation. He underwent excision of cystic SOL under GA. A curved incision was made in the upper part of the swelling then the skin flap raised around the swelling.

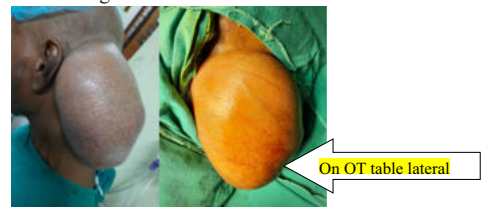


Figure 4- Preoperative photograph.

A soft cystic pinkish to reddish-brown mass exposed. We completely excise the mass. Proper hemostasis secured, skin closed with 2-0 non-absorbable monofilament suture. The weight of the mass was 400mg. The postoperative period was uneventful and he was discharged after 7 days of admission with hemodynamically stable condition.

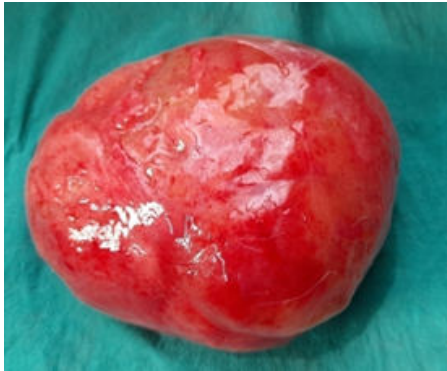


Figure 5. Pinkish to reddish-brown mass after excision of swelling.

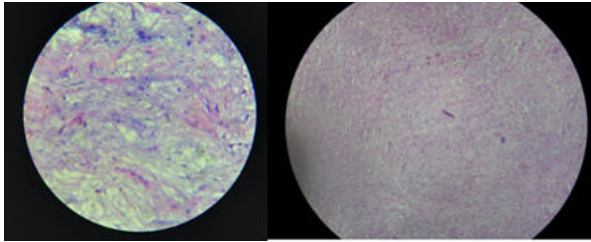


Figure 6. Histologically, the tumour shows a proliferation of wavy spindle-shaped cells embedded in a mucinous stroma (H&E, original magnification 150 X).

3. HISTOPATHOLOGICAL EXAMINATION: Histopathology results revealed grossly single globular greyish-white to greyish-brown tissue piece measuring (14x13x7.5) cm. On cut section, whitish mucinous fluid came out. No variegated appearance was noted and microscopic examination show spindle-shaped cells with wavy nuclei. Mast cells were present and there was abundant mucin (Figure 6).

4. IMMUNOHISTOCHEMISTRY: (MN) positive immunohistochemical staining for S100 protein.

5. FOLLOW-UP AND OUTCOME: The patient is followed up after 12 days and the wound was healthy without any complication and he will be followed up monthly to look for any recurrence till 1 year.



Figure 7. This photograph 12 days after the operation. The wound is healthy without any complication.

6. DISCUSSION :

Myxoid neurofibromas (MN) have been described under many different terms including nerve sheath myxoma, Pacinian neurofibroma, neurothekeoma, bizarre cutaneous neurofibroma, and lobular neuromyxoma. The tumor is most notorious for being a component of the NAME syndrome (nevi, atrial myxoma, myxoid neurofibroma, and epheides) [2]. Myxoid neurofibroma is a benign tumor of perineurial cell origin [3]. Myxoid neurofibroma has a higher incidence in teenagers and young adults [4]. Myxoid neurofibromas are rarer than the typical ones. These tumors are confused often clinically and histopathologically with myxoma. Myxomas have no nerve involvement and are S100 positive and negative for CD 34 [5]. This tumor has a higher incidence in young adults [6]. It usually presents as a non-tender solitary nodule [7]. The most common locations of the MN are the face, shoulders, arms, periungual, and the feet. [8]. To our knowledge, this is the first time that a scalp location has been reported. Myxoid neurofibroma is usually a solitary lesion, as in our patient;

however, they can be numerous and may recur following an incomplete initial excision. Histological examination of myxoid neurofibroma is paramount to exclude malignant tumors such as neurofibrosarcoma and epithelioid sarcoma.

The other differential diagnosis are the following.

- (i) Intramuscular myxoma (IM) has a predilection for extremity with most being located in the proximal thigh, gluteal region, and shoulder girdle.
- (ii) Periungual Myxoid neurofibromas are slow-growing tumors in the fingernail bed. The tumor was elevating and bowing the fingernail.
- (iii) Cellular myxoma showing moderate cellularity (20%), arching blood vessels, and collagenous stroma.
- (iv) Aggressive angiomyxoma are perineal or pelvic in location, shows prominent medium to large vessels.
- (v) Low-grade fibromyxoid sarcoma shows alternating fibrous myxoid patterns with swirling and whorled appearance.
- (vi) Myxoid liposarcoma shows an arborizing vascular pattern, signet ring type of lipoblasts, and is more cellular PDF 1.
- (vii) Ganglioneuroma: A benign tumor of neural crest origin comprised of ganglion cells arising from nerves. Most commonly found in the posterior mediastinum and retroperitoneum. Immunohistochemistry: S100 protein stains Schwann cells and synaptophysin stains ganglion cells.
- (ix) Schwannoma: A benign peripheral nerve sheath tumor comprised predominantly of Schwann cells. Associated with somatic and germline mutations in NF2. Histologically often more cellular, circumscribed, with Verocay bodies and Antoni A and B areas. Immunohistochemistry: Diffuse, uniform staining with S100 [9].

The usual first-line treatment is total excision of tumor [10], but only for cosmetic or diagnostic reasons. Some reports have mentioned the danger of these tumors masquerading as malignancies (mainly neurofibrosarcoma or epithelioid sarcoma) [11]. Histopathological and immunohistochemical stains are useful in the final diagnosis [12].

7. TREATMENT: For most of the cases, complete surgical excision is the preferred treatment, with local recurrence extremely rare [13]. Our patient underwent complete surgical resection without complication. He has no recurrence to date.

8. PROGNOSIS: All types of neurofibromas are benign. Local recurrence is extremely rare complete excision of the lesion, and the risk of malignant transformation is exceedingly low; however, malignant transformation affects approximately 10% of patients with NF1 [14].

9. COMPLICATION: The complications related to localized neurofibromas are mild and typically related to surgical excision of the lesion: pain, bleeding, scarring, and local infection.

10. CONCLUSION: The present case was that of a 56 yrs male patient with large scalp swelling involving the occipital area with extension to the nape of the neck. We excise the total mass. The histopathology of our case was that of myxoid neuroma (MN). The patient is on follow up and has shown no recurrence to date. **This is the first case reported in the literature on myxoid neuroma presenting in the scalp. We report this case because of the rarity of both the tumor and its scalp location and also a giant size and to provide a review of the literature.**

FOOTNOTES:

-SUKANTASIKDAR (@SUKANTASIKDAR3) Twitter.

11. DECLARATION OF COMPETING INTEREST: The authors declare that no conflicts of interest pertinent to this case report.

12. FUNDING: No source of funding to be declared.

13. ETHICAL APPROVAL: No institutional review board is required for the publication of a case report at our institution.

14. PATIENT CONSENT: The written consent was obtained from the patient directly.

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