

Unusual Presentation of a Rare Soft Tissue Neoplasm; Myofibroma of the Ear

Dr. Abhilasha Asthana¹, Dr. Amit Kumar Singh²

¹Senior Lecturer, Dr. B. R. Ambedkar Institute of Dental Sciences and Hospital, Patna – 801503, India

²Senior Resident, IMS, BHU, Varanasi, UP – 221005, India

Abstract: *Myofibroma are benign mesenchymal tumors, commonly found in the dermis and subcutaneous tissues of the head and neck. Myofibromatosis is generally considered to be a benign process, The prognosis is good for patients with a solitary myofibroma or multicentric myofibromatosis without visceral involvement because these lesions often regress spontaneously. Cases of isolated lesions, especially in infancy, expected management is adequate because these lesions typically involute during the first 2 years of life. Chemotherapy is an option for reducing the size of the lesion and alleviating associated symptoms. Recurrence rate for solitary myofibroma is low.*

Keyword: Myofibroma, Myofibromatosis, autosomal - dominant and recessive, desmin and S-100 protein.

1. Case Report

A 15 year old girl presented with the complaints of right post aurial swelling which apparently started as a small swelling 8yrs ago. During her schooling period, she has a history of repeated trauma to the ear. Initially the trauma caused only little bruises and pain which subsided by itself. As time lapsed her mother noticed a small diffuse swelling behind the right ear, which was non tender with normal overlying skin. Prior to this incidence the ear was anatomically normal without any complaints.

After few months the swelling gradually progressed, without any signs of inflammation or any change in overlying skin. There was no history of fever, visual or facial disturbances, ear discharge or bleeding. There was no history of any surgical or medical illness. General and systemic examination revealed no abnormality.

Local examination of right ear revealed a subcutaneous mass in the post aurial region (Fig. 1) measuring 7cm x5cm x3cm. The lesion was soft on palpation, with no fixations to underlying structures. Mass was freely mobile but firmly adherent to the overlying skin, which was normal. The swelling had progressed and grown through the concha of the right ear and was merging with the tragal cartilage (Fig. 2)

CT scan demonstrated a swelling of 6.65 cm x 3.46cm intensely and heterogeneously enhancing soft tissue mass lesion with few areas of necrosis seen external to mastoid bone and external auditory canal on the right extending posteriosuperioly to the right lamdioid suture, bounded inferiorly by right masseter muscle and anteriorly extending to the right superficial parotid gland infiltrating it. There was no intracranial or intratympanic extension. There was a defect of 1.5cm over the cortex in postero superior region of the occipital bone.(Fig. 3)

An incision biopsy was done under GA with a post aurial approach. The swelling was discovered to be immensely vascular soft tissue mass, firmly adherent to the subcutaneous plane. Punch biopsy was taken for

histopathological examination as it was bleeding profusely. Histopathological analysis of the biopsy demonstrated a mass showing intricate mixture of blood vessels and fibrous stroma containing stellate shaped fibroblast. There was vast proliferation of blood vessels size ranging from capillary to venous size. There was a sparse infiltration by mononuclear cells predominantly lymphocytes and plasma cells mixed with mature adipose tissue. No evidence of malignancy was observed. Histopathological Impression was: Myofibroma (Fig. 4)

Two weeks later keeping 2 units of whole blood reserved the patient was posted for excision of the total mass which was completely excised even though excessive bleeding was observed, by maintaining all parameters it was successful and uneventful for 24 hours. Post operative result was uneventful without any recurrence or residual region. Treatment with systemic antibiotics was given for 7 days along with haematinics. After follow-up of 3 months the surgical wound was healed.

2. Discussion

Myofibroma are benign mesenchymal tumors that are commonly found in the dermis and subcutaneous tissues of the head and neck. First described by Stout (2) in 1954 and further classified by Chung and Enzinger (1) in 1981. Myofibroma is characterized histologically by a proliferation of fibroblasts and myofibroblasts. Immunohistochemical stains are positive for smooth – muscle actin and vimentin and negative for desmin and S-100 protein. Myofibromatosis is generally considered to be a benign process, unlike aggressive fibromatosis and fibrosarcomas, which are invasive lesions with metastatic potential. Nonetheless, bony destruction may occur in myofibromatosis, and some authors have described aggressive and infiltrative lesion behavior (3).

Types of myofibroma

There are three types of myofibroma:

- Solitary myofibroma,
- Multicentric fibromatosis without visceral involvement,
- Multicentric fibromatosis with visceral involvement.

The prognosis is good for patients with a solitary myofibroma or multicentric myofibromatosis without visceral involvement because these lesions often regress spontaneously. Conversely, myofibromatosis with visceral involvement can be fatal within days or weeks of birth, usually as a result of pulmonary or gastrointestinal involvement (4). A positive family history is often present in these cases, although both autosomal - dominant and autosomal - recessive transmission have been described. Solitary nodules and diffuse myofibromatosis usually occur in the head and neck, and there is a male preponderance (4, 8). Superficial tumors usually present as palpable, rubbery, firm nodules that are freely mobile, while deeper lesions are typically fixed. Overlying skin changes, such as a purplish discoloration similar to a hemangioma, may be present. Ulceration and skin atrophy have also been described (9).

The nonspecific nature of the clinical presentation and the relative rarity of myofibroma present a diagnostic challenge. Even when a pathologic specimen has been obtained, these lesions continue to elude diagnosis because many other lesions display areas of myofibroblastic cells. Other cutaneous and subcutaneous lesions that are considered in the differential diagnosis include leiomyoma, hemangiopericytoma, and fibrous hamartoma of infancy, cutaneous inflammatory pseudotumor, desmoid tumor, nodular fasciitis, plexiform fibrohistiocytic tumor, neurofibroma, and dermatomyofibroma (9, 10).

Our case was unusual in respect to age of presentation, location and a possible relation to trauma. A review of the literature identified only a small number of cases of solitary myofibroma that involved the cheek or face. Only 4 other cases of isolated myofibroma of the cheek or face have been reported (3, 5, 6, 13, 17). Most cases of myofibroma occur in infants; presentation in older children and in adults is rare.

A history of trauma is not typically associated with myofibroma. It is interesting that our patient reported a history of minor blunt trauma to the site 8 yrs prior to presentation. This raises the possibility that her myofibroblastic lesion represented an exuberant reactive process. On the other hand, it is also possible that the trauma was a coincidental occurrence that merely drew attention to a pre - existing myofibromatous lesion. Also, the possibility that a myofibroma could occur as a reaction to local trauma would not explain the multiple lesions that are seen in patients with diffuse myofibromatosis. There is evidence to suggest that myofibroblasts also play a role in scar formation. (15) The histologic similarity between myofibroma and scar tissue suggests that myofibroma may be the result of a self - limited reactive process rather than true neoplasm.

3. Diagnosis

Myofibroma present an ongoing diagnostic challenge. Routine radiological imaging is often non diagnostic and findings can differ depending on the location of the lesion. Solid - tissue lesions are difficult to distinguish from other mesenchymal tumors. Other imaging modalities may help

narrow the differential diagnosis. Ultrasonography of such a lesion may yield a hypoechoic region with or without a cystic space. Magnetic resonance imaging typically demonstrates isodensity to muscle on non contrast T1-weighted imaging and hyper intensity on T2- weighted imaging. (16) Additionally, myofibroma with increased vascularity enhance with gadolinium. (16) Even with radiological evidence, biopsy is ultimately necessary to make a definitive diagnosis and to rule out a more aggressive neoplasm.

4. Management

The management of myofibroma is not standardized. The appropriate treatment varies according to the clinical situation. In cases of isolated lesions, especially in infancy, expectant management is adequate because these lesions typically involute during the first 2 years of life. We performed an Incisional biopsy after our patient had undergone CT, and 1 month of inpatient observation. We were able to excise most of the lesion, but a complete excision was precluded because of the potential risk of excessive blood loss and lack of exact demarcation of the mass while operating.

A conservative management strategy that is limited to obtaining a diagnostic biopsy sample is considered to be sufficient unless the lesion has caused a functional impairment or cosmetic deformity. Complete surgical excision should be undertaken in cases of symptomatic visceral lesions. In some cases, intimate involvement of vital structures may dictate a more conservative excision. In cases where surgery might cause major morbidity, chemotherapy is an option for reducing the size of the lesion and alleviating associated symptoms. Recurrence rate for solitary myofibroma are low. Risk factors for recurrence include a location on the extremities, age greater than 5 years at presentation, and a previous incomplete excision. (17).

References

- [1] Chung EB, Enzinger FM. Infantile Myofibromatosis. *Cancer* 1981; 48:1807-18.
- [2] Stout AP. Juvenile fibromatosis. *Cancer* 1954;7:953-78
- [3] Beck JC Devaney KO, Weatherly RA, et al. pediatric myofibromatosis of the head and neck. 1999; 125:39-44.
- [4] Fibrous tumor of infancy and childhood. In: Enzinger FM. Weiss SW. soft tissue tumors. 3rd ed. St Louis Mosby, 1995;357-63
- [5] Sleeman DJ Eveson solitary infantile myofibromatosis. *Br J Oral Maxillofacial surg.* 1991;29:277-8
- [6] Guitar J, Ritter JH. Wick MR solitary cutaneous myofibroma in adults: report of 6 cases and discussion of differential diagnosis. *J Cutan Pathol* 1996; 23:437-44.
- [7] Wolfe JT 111, Cooper PH, solitary cutaneous "infantile" myofibroma in 49yrs old woman. *Hum Pathol* 1990; 21:562-4.
- [8] Shields CL, Husson M, solitary intraosseous infantile myofibroma of the orbit roof. 1998; 116:1528-30.

- [9] Wyatt AJ, Hansen RC. Pediatric skin tumors. PCNA 2000; 47:937-63.
- [10] Hogan SK Salassa JR. recurrent adult myofibromatosis. Case report. Am Jr Clin Pathol 1992; 97:810-14
- [11] Walsh RM, Leen EJ, Gleeson MJ. Solitary infantile and adult myofibromatosis of nasal cavity. a report of 2 cases. JLO 1996; 110:574-7.
- [12] Balakirshnan, Pujary, Shah, Hazarika, solitary adult myofibroma of the pinna . JLO 1999; 113:155-7.
- [13] Feltcher CD, Achu P, Van Noorden S. infantile myofibromatosis, histochemical and immunohistochemical study suggesting true smooth muscle differentiation. Histopath 1987; 11:245-58.
- [14] Smith KJ, Skeleton HG, Barret TL et al. cutaneous myofibroma Mod Pathol 1989;2:603-9
- [15] Terries DJ. Dynamics of wound healing, In Bailey BJ, ed atlas of head and neck surgery otolaryngology 2nd ed. 1996:223-32.
- [16] Kayes AV, Bancroft LW. Tenneson GS, myofibroma of upper arm in 52 yrs old woman. Skeletal radiol. 2002; 31:240-5.
- [17] Ashutosh Kacker, MD, Department of Otolaryngology—Head and Neck Surgery, New York Presbyterian Hospital—Weill Medical College of Cornell University, New York City



Figure 1



Figure 2

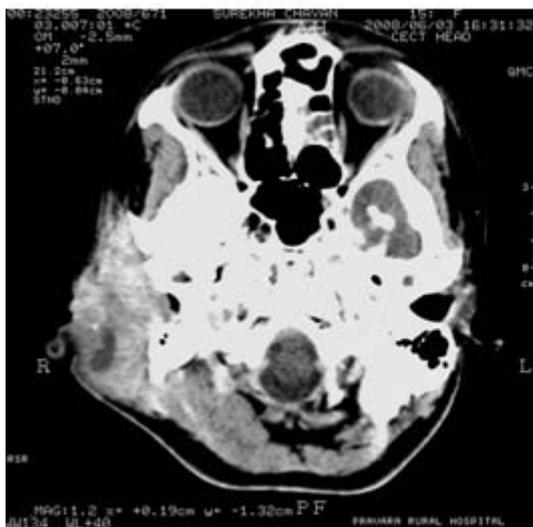


Figure 3

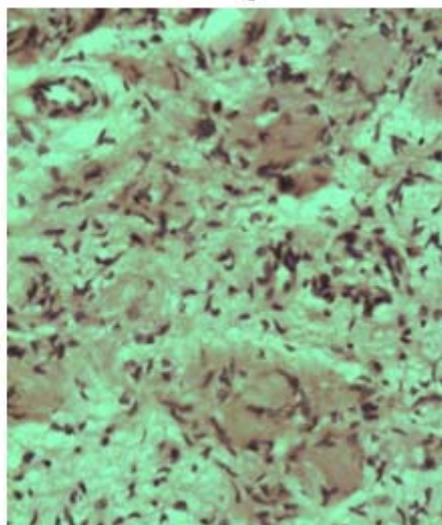


Figure 4

- Figure 1:** Post Aural Soft Tissue Swelling.
- Figure 2:** Swelling Merged with the Conchal Cartilage
- Figure 3:** CT Scan of the Soft Tissue Swelling
- Figure 4:** Histopathology of the Excised Specimen