ISSN : 2350-0743

www.ijramr.com

*International Journal of Recent Advances in Multidisciplinary Research*

*Vol. 08, Issue 07, pp. 7008-7010, July, 2021*

# RESEARCH ARTICLE

# EXTRAMUSCULAR MYXOMA- BEING AWARE OF THE AGGRESSIVE LOOKING RARE LESION

**Dr. Navin Kasliwal M.S.,[1,*] Dr. Nilesh Chandak M.Ch and [2]Dr. Sonika Ranganath**

[1]Surgical Oncologist and Assistant Professor, MGM medical college, Aurangabad
[2]Surgical Oncologist and Associate Professor, UPMC, Jalgaon
[3]Surgical Resident, MGM medical college, Aurangabad

**ARTICLE INFO**

***Article History:***

Received 25th April, 2021
Received in revised form
14th May, 2021
Accepted 10th June, 2021
Published online 30th July, 2021

***Keywords:***

Extramascular Myxoma,
Myxoma, Surgery.

**ABSTRACT**

Extramuscular soft tissue myxomas are extremely rare with only 4 documented cases in english literature till date. Myxomas are benign connective tissue tumours of unclear histogenesis and etiology. Virchow coined the term Myxoma in 1871 while Stout defined the criteria for diagnosis in 1948. Cardiac Myxomas are the most commonly reported and researched, and surgery is the accepted treatment modality. Extra cardiac myxomas are less common and predominantly occur in the head neck region. We review the literature about soft tissue myxomas in the light of a case of an Extramuscular myxoma in a 1 yr old kid. We propose inclusion of myxomas in differential diagnosis of neck masses and recommend surgery in the form of complete excision as the primary modality for treatment in these tumors though the clinical and radiological picture is deceptive. Paucity of available literature and rarity of these tumors demands vigilant evaluation to frame guidelines for their appropriate management.

## INTRODUCTION

Extramascular myxomas are a very rare type of extracardiac soft tissue myxomas with very few cases reported in literature. The paucity of clinical occurrence and available literature about this rare entity explains the unavailability of clear guidelines for its management. In the light of a treated patient with a 3 year follow up now, we review the literature available about extramascular myxomas in an attempt to formulate the working guidelines for the management of the same.

**The case details:** A 1 year male child presented with a large mass in the left side of neck in the upper half. The mass had restricted side to side mobility and pushed the trachea to the right. It was a firm non tender mass with a history of gradual increase in size over 4 months. The kid had nodysphagia or change of voice. Growth parameters were normal with no physical or mental developmental lag. Contrast CT scan was done which showed a 8 x 11 cm mass in the left neck, free from the muscles, pushing the trachea larynx and esophagus anteriorly and to the right. It was abutting the prevertebral fascia posteriorly. The neurovascular bundle was pushed but not invaded. Superiorly the mass was reaching the level of mastoid on the left.

***Corresponding author: *Dr. Navin Kasliwal M.S.*,**
Surgical Oncologist and Assistant Professor, MGM medical college, Aurangabad.

Under general anaesthesia a utility apron neck incision was taken. Ryles tube was placed upfront to identify the esophagus. Subplatysmal flaps were raised. The neurovascular bundle was defined and looped. The mass was gradually dissected and excised en masse after lateralisation of the trachea and the esophagus. The left recurrent nerve was identified and preserved. Post operative recovery was uneventful. There was no evidence of neurovascular deficit. Swallowing and breathing were normal. Patient was started back on orals in 6 hours and discharged after 4 days. The kid is disease free and on regular followup and it is now three years from surgery.

## DISCUSSION

Myxomas are benign connective tissue tumours. They are locally infiltrative, benign lesions. These mucinous tumours have a specific consistency resembling an umbilical cord in pathological appearance: globule-like, sometimes on a peduncle but usually sessile with wide attachment at base. In 1871, Virchow coined the term Myxoma when he observed tumors similar to the umbilical cord mucinous tissue on histology.[1]

**Definition:** Stout, in 1948, reported 49 patients with myxoma and devised criteria for the diagnosis of myxoma, stating that they: [2]

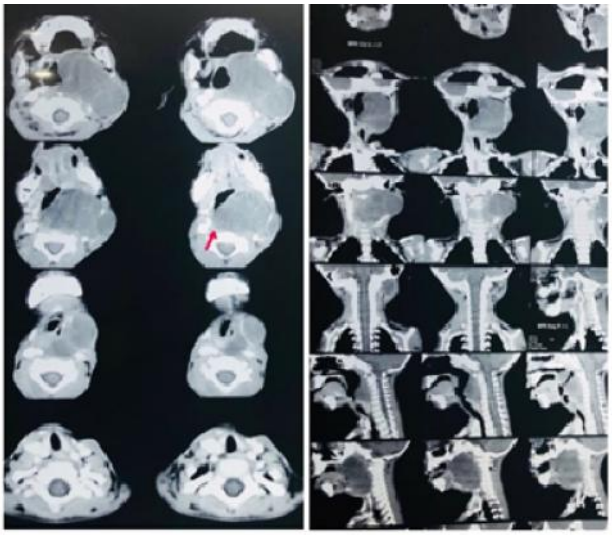

**Figure 1. CECT of neck showing the mass arising from left side of the neck and pushing the trachea, larynx, esophagus and neurovascular bundle anteriorly and right**

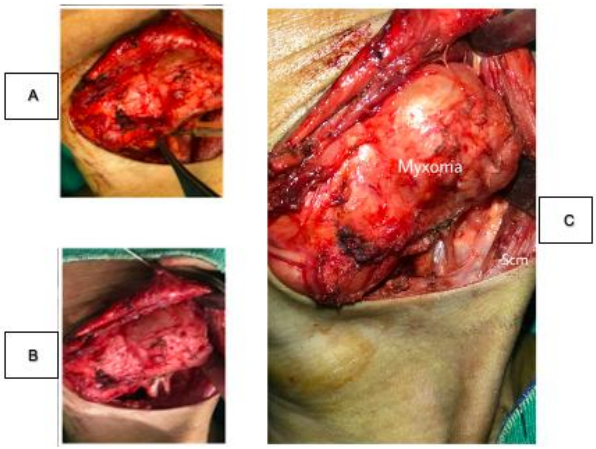


**Figure 2. Dissection anatomy**

- are trueneoplasms- composed of stellate-shaped cells in loose mucoid stroma;
- have no recognisable cellular elements and specifically no chondroblasts, lipoblasts or rhabdomyoblasts;
- are poorly vascularised;
- are of unicentric origin and
- displaybenign behaviour.

Myxoma is the most common intracardiac (atrial) tumor. Extracardiac myxomas are less common and occur in aponeurotic tissues, bone, genitourinary tract, skin, retroperitoneum, intestine, pharynx, joints and skeletal muscles.[4]

Myxomas are of several types depending on the organ where they develop, and their aggressiveness.[7,10]

- Cutaneous myxoma (on skin or subcutaneous tissue)
- Atrial myxoma (the most common primary heart tumors, usually located in the left atrium),
- Intramuscular myxoma (in skeletal muscles; very rare).
- Juxta-articular myxoma (shoulder, ankle or hip),
- Digital myxoma (tip of the fingers).
- Aggressive angiomyxoma (vulvovagina, peritoneum),
- Angiomyofibroblastoma (well marginated- skin or vulva),
- Nerve sheath myxoma (develops at the layers of the nerve -legs, knees),
- Pseudomyxoma peritonei (aggressive, spreads quickly over the peritoneum).

Myxomas appear more frequently in women than men for unknown reasons.[9]The etiology of myxomas remains elusive. Some authors believe trauma to be a cause of formation of myxomas.[10] Another proposed theory is the growth of polysaccharide-producing cells in the neoplastic process. About 10% of myxomas are genetically influenced while the rest are sporadic.Examples of inherited syndromes that include myxomatous tumours are [8-10]

- Carney complex (multiple myxomatous tumours, skin hyperpigmentation, schwannomas, testicular tumours and pituitary adenoma) and
- Mazabraud syndrome (myxomatous tumours of skin, heart and bones associated with McCune-Albright syndrome).

Extracardiac myxomas are benign tumors with a slow growth rate and low mitotic activity; they usually present in the 4th to 6th decades of life more in women. These most commonly occur in the head and neck region in skin tissue, the jaw bones and maxillary and soft tissues of the face. Most of them are of osseous origin or arise intramuscular, surrounded by skeletal muscles. Though head neck osseous myxomas are common, mostly in the maxilla and mandible, cases of myxoma of peripheral bone (osseous) have also been reported in the literature. Amongst the initial cases, McClure and Dahlin first reported three. Further myxoma of the phalanx of the toe was reported in 1978 and a case of periosteal myxoma of the femur reported in 1972. Soft tissue myxomas are also rare lesions again commonly occuring in head and neck area. These are usually intramuscular. Enzinger, in 1965, identified characteristic properties of a myxoma removed from intramuscular tissue, and described it as intramuscular myxoma.[8] Intramuscular myxomas can occur in the muscles of the thigh, buttocks, shoulder and upper extremities, in patients between 40 and 70 years of age, with female predominance.[6,11] They are asymptomatic except for a slow growing mass. Most of the extracardiac soft tissue myxomas are osseous or intramuscular, Intramuscular myxoma usually occurs as an isolated lesion in form of a painless palpable mass. The first case of intramuscular myxoma was described in 1965. Extramuscular myxomas of the neck are extremely rare. Only 4 cases of extramuscular myxoma have been reported in literature till date.[7,12].These are aggressive looking tumors on imaging and on clinical evaluation and may not be considered for surgery if only the clinical and radiological picture is considered. Myxomas and fibromyxomas have very similar clinical, radiological and biological features. Both types are benign lesions without the potential to metastasise and it is not of major importance to differentiate since both have similar treatment protocol and recurrence rates, it is important to differentiate these entities from fibrous dysplasia because of the higher recurrence rate of fibrous dysplasia. Clinical evaluation to assess the extent of disease and cross sectional imaging (CT scan) followed by a needle biopsy is needed for evaluation. The appropriate treatment for all myxomas is surgical excision and almost all tumors completely resected show a very favorable outcome.

The high recurrence rate previously reported is probably due to incomplete excision (e.g. when enucleation or curettage is used in osseous lesions). The other differential diagnoses include other myxoid neoplasms and proliferative lesions of the soft tissue. Benign lesions such as myxolipoma, myxoid neurofibroma, neurothecoma, myxochondroma and nodular fasciitis and different types of low grade myxoid sarcomas should also be considered.[11,12] Also to consider are low grade myxofibrosarcoma, myxoid liposarcoma, extraskeletal myxoid chondrosarcoma and low grade fibromyxoid sarcomas.[12] Also, extramuscular myxoma differential in the cervical region must be made with the most common cystic tumors of the neck, such as the branchiogenic cyst, laryngocele, spinocellular carcinoma cystic metastasis and thyroid papilliferous carcinoma cystic metastasis.[7] Treatment of solitary myxomas is wide surgical excision. No metastasis, recurrence or malignant change has been reported with adequate surgical excision. However, recurrence has been reported in a small number of patients undergoing enucleation and incomplete resection in some cases of intramuscular or osseous myxomas.

**Conclusion**

**Extramascular soft tissue myxomas are**

- Rare
- Benign
- Usually occur in neck
- Have a locally aggressive clinical picture
- Look extensive and push but do not infiltrate nearby structures on imaging.
- Need surgical excision - only.
- Good prognosis with rare recurrence and no metastases.

We recommend keeping the possibility of a extramascularmyxoma in mind while evaluating neck soft tissue lesions so that curative surgical excision is contemplated in spite of aggressive and deceptive clinical and radiological picture.

## REFERENCES


1. Virchow R. Die cellularpathologie in ihrerBeegrundung auf physiologische and pathologischeGewebelehre. Berlin, Germany, Verlag von August Hirschwald. 1871, 563.
2. Stout AP. Myxoma, the tumor of primitive mesenchyme. Ann Surg. 1948, 127:706-719.
3. Luna A, Martinez S, Bossen E: Magnetic resonance imaging of intramuscular myxoma with histological comparison and a review of the literature. Skeletal Radiol. 2005, 34: 19-28
4. Andrews T, Kountakisse, Maillard AA. Myxomas of the head and neck. Am J Otolariyngol. 2000, 21:184-9.
5. Galera-Ruiz H, Martin-Gomez R, Esteban-Ortega F, Congregado-Loscertales M, Garcia Escudero A. Extramuscular soft-tissue myxoma of the lateral neck. Rev LaryngolOtolRhinol. 2001, 122:259-61.
6. Fletcher CDM, Unni KK, Mertens F (Eds). World Health Organization Classification of Tumors. Pathology and Genetics of tumors of Soft tissue and Bone. IARC Press: Lyon, 2002.
7. Melo GM, Tavares TV, Curado TAF, Cherobin GB, Gonçalves GNH, Ribeiro CMF, et al. Myxoma of Cervical Soft Tissue: Case Report and Literature Review. Int. Arch. Otorhinolaryngol. 2008;12(4):587-590
8. Enziger FM. Intramuscular myxoma. Am J Clin Pathol. 1965, 43:104-13.
9. Ghosh BC, Huvos AG, Gerald FP, et al. Myxoma of the jawbones. Cancer. 1973, 3:237-40.
10. Allen PW. Myxoma is not a single entity: a review of the concept of myxoma. Ann Diagn Pathol. 2000, 4:99-123.
11. Papadopoulos EJ, Cohen PR, Hebert AA. Neurothekoma: report of a case in an infant and review of the literature. J Am Acad Dermatol. 2004, 50:129-34.
12. Daniel et al.; Giant Fungating Cutaneous Myxoma of the Head and Neck: An Unusual Presentation JCTI, 9(4): 1-5, 2019


*******