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GENETICS AND MOLECULAR INSIGHT INTO INFLAMMATORY MYOFIBROBLASTIC TUMOUR OF ORAL CAVITY – A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Introduction: Inflammatory myofibroblastic tumour (IMT) is a rare benign mesenchymal neoplasm composed of myofibroblastic spindle cells and inflammatory infiltrate. First described in the lung by Brunn in 1939, it is uncommon in the oral cavity, with only 58 reported cases to date. IMT frequently affects children and young adults and may mimic malignancies clinically and radiographically due to its potential for vascular invasion, aggressive progression, and local recurrence. While its etiology remains unclear, factors such as trauma, Epstein-Barr virus, and ALK gene rearrangements have been implicated. ALK positivity, detected via immunohistochemistry or FISH, is a distinguishing feature in up to 60% of cases.

Case Presentation: A 9-year-old female presenting with a 6-month history of progressively enlarging swelling in the right buccal mucosa following facial trauma. Clinical and radiological evaluations suggested a neoplastic etiology, and excisional biopsy revealed myofibroblastic spindle cell proliferation with inflammatory infiltrate, confirming IMT.

Management and Prognosis: The tumour was surgically excised. The patient is under regular follow up.

Conclusion: IMT's pose a diagnostic challenge because of histopathological overlap with other fibroblastic and myofibroblastic lesions. High recurrence rate necessitates long-term follow-up.

KEYWORDS: ALK positivity, Epstein Barr virus, FISH, Immunohistochemistry, Inflammatory myofibroblastic tumour.

INTRODUCTION

Inflammatory myofibroblastic tumour (IMT) is a rare benign mesenchymal neoplasm characterized by a myofibroblastic spindle cells interspersed with an inflammatory infiltrate. Brunn in 1939 was the first to observe inflammatory myofibroblastic tumour in the lung. Liston et al in 1981 was the first to report a case of inflammatory myofibroblastic tumour in the oral cavity[1]. Various nomenclature such as inflammatory

pseudotumour, plasma cell pseudotumour, inflammatory myofibrohistiocytic proliferatio, inflammatory fibrosarcoma, plasma cell granuloma and pseudosarcomatous lesion are all synonyms that have been used to describe inflammatory myofibroblastic tumour because of varying histologic appearance. The most common and recently used term now is the inflammatory myofibroblastic tumour[2]. WHO defined inflammatory myofibroblastic tumour as a distinctive neoplasm composed of fibroblastic and myofibroblastic spindle cells accompanied by inflammatory infiltrate of lymphocytes, plasma cells and eosinophils. Lung is the commonest site being affected followed by gastro intestinal system. Occurrence in the head and neck region is rare with a incidence of 15%. In the maxillofacial region larynx is the most common site affected[3]. Its presence in oral cavity is exceedingly uncommon with only 58 cases reported to date. Although various pathogenesis have been suggested the etiology of IMT still remains unclear. Trauma, exposure to radiotherapy, association with Epstein Barr virus, ALK gene have been identified as possible causes[4]. Its incidence is rare and affects children and young adults more frequently with a prevalence ranging from 0.04% to 0.7% worldwide, irrespective of gender and race.

IMT may mimic a malignant neoplasm both clinically and radiogarphically. It has a potential for vascular invasion, aggressive progression and local recurrence[5]. Due to its unclear clinical presentation IMT needs to be differentiated from other granulomatous, infectious, autoimmune and malignant lesion on the basis of histopathologic examination and immunohistochemical analysis. Although IMT is uncommon in the maxillofacial region, precise diagnosis is crucial to differentiate it from more severe differential diagnoses and to ensure proper treatment.

This case report presents a case of IMT in a 9 year old female child.

CASE PRESENTATION

A 9 year old female patient reported to the outpatient department with the presence of swelling in the right buccal mucosa. There was no relevant past medical and dental history. There was a history of fall 6 months backs with sustained injury to the right side of the face. The lesion was of six months duration and was progressively enlarging. Extraoral examination revealed a well defined swelling with facial asymmetry (Fig 1). Intraoral examination revealed well circumscribed, pedenculated nodule of size 2.5×1.5 cm in diameter approximately. The colour of the lesion was as same as the surrounding normal mucosa. Ultrasonography revealed well defined heteroechoic solid in the subcutaneous and intramuscular plane in the right buccal mucosal region measuring around 2.5×1.5 cm showed internal vascularity on colour Doppler suggestive of neoplastic etiology (Fig 2). Fine needle aspiration cytology showed blood mixed aspirate macroscopically, scanty cellular and fragments of fibroconnective tissue with RBC's in the background microscopically. The differential diagnosis was made as minor salivary gland pathology. An excisional biopsy was done and the specimen was sent for histopathological examination. Gross examination revealed that the specimen was firm in consistency, greyish white in colour, measured around 4×4 cm in diameter approximately (Fig 3). Microscopic examination showed a mixture of irregularly arranged fascicles of spindle cells and inflammatory cells. The spindle cells appear bland to mildly atypical. The connective tissue stroma showed diffuse and focal infiltration of neutrophils, lymphocytes, plasma cells and histiocytes (Fig 4 & 5). The background stroma showed myxoid foci. The final diagnosis was given as inflammatory myofibroblastic tumour. The tumour was surgically resected. No recurrence was observed after 1 year follow up.

DISCUSSION

Myofibroblasts are ubiquitous cells showing ultrastructural features of both fibroblasts and muscle cells. IMT is a rare controversial entity the etiology of which still remains enigmatic. WHO describes IMT as a distinctive neoplasm composed of myofibroblastic cells admixed with inflammatory infiltrate. Several risk factors have been identified including minor trauma, smoking, IgG4 related disease[6]. Some hypotheses propose that an abnormal immune response to antigens or viruses such as Human Herpes Virus 8 and Epstein Barr Virus, may play a role. In view of the absence of the immunohistochemical profile and variable phenotype, the diagnosis of IMT has traditionally been a diagnosis of exclusion, with a wide differential diagnosis that includes conditions ranging from local inflammatory process to inflammatory fibrosarcoma. It has been well established that in up to 60% of IMT ALK rearrangements occur. ALK is paired with a broad range of 5' fusion partners which all result in aberrant ALK homodimerization in the absence of ligand and constitutive activation. TIMP3 (~50%) was the most frequent fusion partner in the head and neck series. The gold standard for detection of ALK rearrangements is by FISH or NGS. Immunohistochemistry show cytoplasmic positivity for ALK[7].

IMT often present with site specific symptoms. Oral IMT often present as slow growing, painless, well circumscribed nodular mass. IMT have a non specific radiographic presentation. Ultrasound may reveal a hypoechoic or hyperechoic mass with well or ill defined borders with increased vascularity on Doppler studies. Magnetic resonance imaging show low signal intensity on T1 and T2 weighed images with restricted diffusion due to fibrosis. CT imaging shows heterogenous density in unenhanced CT images in larger masses and homogenous density in unenhanced CT images in smaller masses[8].

Biologically elevation of erythrocyte sedimentation rate and c-reactive protein, hypochromic normocytic anemia, thrombocytosis and hypergammaglobulinemia is seen in patients with IMT due to induction of inflammation[9].

A systematic review of previously reported cases of IMT was performed. A systematic search of the literature was conducted in PubMed, Scopus, and Google Scholar using the keywords "Inflammatory myofibroblastic tumour," "IHC," "ALK positivity." Only English-language case reports and case series with adequate clinicopathological data were included. A total of 58 studies reporting cases of IMT have been reported. Table 1 illustrates a review of literature of IMT from 1981 to 2024 including the present case.

It was seen that lesions occur over the wide range of age group from 2 to 82 years with slight female predilection. The lesions often present as a firm, indurated swelling. Buccal mucosa is the most common site affected in the oral cavity.

Microscopically, myofibroblastic spindle cell proliferation with mixed inflammation is seen. Variety of histological pattern such as

1. Spindle to stellate cells admixed with inflammatory cells in a loosely arranged myxoid or hyaline stroma (nodular fasciitis – like).
2. Elongated spindle cells lacking cytologic atypia or overt hyperchromasia with prominent lymphoplasmacytic infiltrate arranged in a fascicular or storiform growth pattern.
3. Hypocellular, scar like pattern with occasional calcifications or metaplastic bone formation is seen.

Ganglion like cells is seen in some tumours. Epithelioid variant is composed of plump round to epithelioid cells with vesicular chromatin, large prominent nucleoli, and amphiphilic to eosinophilic cytoplasm admixed with prominent neutrophilic component in a abundant myxoid stroma[10].

Cytoplasmic positivity for ALK is seen in immunuhistochemistry which is useful in differentiating IMT from other potential mimicking conditions in the head and neck region. Nuclear membrane or perinuclear pattern of ALK staining is seen in epithelioid variant[11]. Table 2 summarizes the differential diagnosis of IMT with a key differentiating feature.

Surgery remains the treatment of choice for localized tumour. En bloc resection with R0 margins is the standardized surgical procedure. Mohs microsurgery or wide local excision involving removal of sarcoma with some surrounding normal tissue is done to ensure complete excision. Advanced or non operable cases are treated with chemotherapy. There is no standard chemotherapy regimen established for IMT. Anthracyclin based chemotherapy with methotrexate ± vinorelbine/vinblastine (MTX-V) is the commonly used regimen. Other regimens include oral cyclophosphamide and docetaxel/gemcitabine. Targeted therapy such as use of ALK inhibitors namely crizotinib[12].

CONCLUSION

Oral IMT is a rare locally aggressive benign mesenchymal neoplasm with non specific clinical, radiographic and pathologic features. It poses a significant diagnostic challenge because of the similar histopathological features with other myofibroblastic and fibroblastic tumour. Diagnosis is difficult with small incisional biopsy. A systematic approach sufficient tumour sampling are essential for establishing a diagnosis. Follow up is crucial to prevent the recurrence due to aggressive nature of this neoplasm.

ACKNOWLEDGMENT AND DISCLOSURE STATEMENT

The authors report no conflicts of interest related to this study.

CONSENT FOR PUBLICATION

Written Informed consent was obtained from the patient for the publication of the images.

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Table 1-Review of literature from 1981 to 2024

Author/ Year	Age/Gender	Location	Radiographic findings	Histopathological features	Final diagnosis	Follow up With no signs of recurrence
Listonet et al., / 1981[13]	4/F	Buccal mucosa	NA	Loose myxoid stroma containing plump fibroblasts and dense eosinophillic bands of collagen	IMT	6 months
	2/F	Buccal mucosa	NA	Stellate appearance with mixture of fibroblasts and chronic inflammatory cells	IMT	10 months
	6/M	Buccal mucosa	NA	Plump and spindle shaped fibroblast in a myxoid stroma with inflammatory cells	IMT	NA
Earl et al., / 1993[14]	44/M	Buccal mucosa	Soft tissue swelling	Plump spindle cells in a fibromyxoid stroma that contained numerous inflammatory cells	IMT	2 years
Ramachandraet et al., / 1995[15]	77/F	Buccal mucosa	NA	Fibroblastic and myofibroblastic spindle cells with aggregates of lymphocytes	IMT	NA

Shek et al., / 1996[16]	20/M 36/F	Buccal mucosa Maxilla	MRI – Infiltrative lesion invading skin and parotid duct CT – Cystic lesion	Fascicles of elongated spindle cells in a mixed inflammatory background Fascicles of plump spindle cells with abundant plasma cells and lymphocytes	IMT IMT	13 months 13 months
Ide et al., / 1998[17]	43/F	Retromolar gingiva	NA	Pseudosarcomatous proliferation of spindle and rounded cells in fascicles admixed with chronic inflammatory cells	IMT	1 year
Cable et al., / 2000[18]	29/F	Hard palate	CT – Small area of ill defined soft tissue lesion	Sheet like proliferation of spindle shaped cells intermixed with variable number of inflammatory cells	IMT	1 year
Ide et al., / 2000[19]	27/M	Tongue	NA	Cellular spindle cell proliferation with chronic inflammatory cell infiltrate	IMT	NA
Pankaj et al., / 2001[20]	-	Tongue	NA	Spindle proliferation in a myxoid stroma	IMT	NA
Jordan et al., / 2003[21]	23/M	Mandible	NA	Spindle cell proliferation with pseudosarcomatous appearance infiltrated with inflammatory cells	IMT	NA

Fang et al., / 2004[22]	23/M	Retromolar gingiva	Radiolocent osteolytic lesion	Spindle cell proliferation with a background of extensive inflammation	IMT	6 months
Brooks et al., / 2004[23]	82/F	Mandible	Soft tissue image of the lesion overlying a resorptive defect.	Proliferation of spindle cells admixed with numerous acute and chronic inflammatory cells	IMT	18 months
Poh et al., / 2004[24]	42/F	Mandible	Well demarcated radiolucent lesion	Short spindle cells with uniformly distributed inflammatory cells in a feather like background	IMT	6 months
Brooks et al., / 2005[25]	82/F	Mandible	Panograhy – Homogenous soft tissue mass	Proliferation of spindle cells admixed with numerous acute and chronic inflammatory cells	IMT	18 months
Gleizal et al., / 2006[26]	23/F	Tongue	NA	Proliferating myofibroblastic cells with plasmocytes and lymphocytes	IMT	14 years
Deshinger et al., / 2007[27]	30/F	Maxilla	CT – Ill defined and heterogenously enhancing mass	Fasicles of spindle shaped cells intermixed with inflammatory infiltrate	IMT	NA
Johann et al., / 2008[28]	33/M	Mandible	NA	Fasicular proliferation of spindle cells with diffuse inflammatory cell infiltrate	IMT	28 months
Oh et al., / 2008[29]	20/F	Mandible	Ill defined osteolytic lesion	Non specific acute and chronic inflammatory cells including plasmacytic hyperplasia with spindle cells	IMT	22 months
Xavier et al., / 2009[30]	23/F	Floor of the mouth	Insignificant	Cellular lesion with focal fascicular arrangement of spindle cells with prominent inflammatory component	IMT	2 years
Eley et al., / 2010[31]	29/M	Maxilla	Soft tissue mass	Spindle cells and stellate cells in a stroma.	IMT	6 years

Satomiet et al., / 2010[32]	14/F	Mandible	Panography – Homogenous soft tissue mass	Haphazard proliferation of fibroblastic and myofibroblastic spindle cells in a inflamed stroma	IMT	10 years
Salgueiredo et al., / 2011[33]	31/F 31/F 60/M	Tongue Alveolar mucosa Buccal mucosa	NA NA NA	Spindled myofibroblastic cells accompanied by an inflammatory cells	IMT IMT IMT	18-48 months 18-48 months 18-48 months
Palaskar et al / 2011[34]	19/M	Mandible	NA	Non encapsulated mass of connective tissue stroma with diffuse and focal infiltration of inflammatory cells with spindle cells interspersed in between	IMT	6 months
Binmandi et al., / 2011[35]	40/F	Gingiva	Unremarkable	Proliferation of spindle cells admixed with numerous chronic inflammatory cells in a fibrous and myxoid backgroung	IMT	4 months
Kim et al., / 2011[36]	24/M	Maxillary sinus	CT – Soft tissue mass involving left maxillary sinus	Plump myofibroblasts and moderate lymphocytes and plasma cells infiltrate	IMT	15 months
Lourenco et al., / 2012[37]	14/M	Tongue	NA	Proliferation of plump spindle cells on a myxoid background	IMT	1 year

Rautava et et al., / 2013[38]	11/F	Maxilla	CBCT – Cystic, osteolytic area	Haphazardly arranged myofibroblastic spindle cells in a inflamed stroma	IMT	3 years
Sah et al., / 2013[39]	30/M	Mandible	CT – Hypodense area	Fasicles of spindle shaped cells admixed with inflammatory cells	IMT	7 months
Lazaridou et et al., / 2013[40]	75/F	Buccal mucosa	CT – Non homogenous mass with moderate enhancement	Neoplastic spindle cells arranged in a storiform and fasicular pattern with variable amount of inflammatory cells	IMT	1 year
Ekici et al., / 2013[41]	75/M	Tongue	NA	Spindle shaped pleomorphic cellular elements in a granulation like tissue	IMT	1 year
Rahman et et al., / 2014[42]	36/M	Maxilla	CT – Heterogenously enhancing soft tissue mass lesion	Low grade collagenous and focally myxoid spindle cell neoplasm	IMT	1 year
Stringer et al., / 2014[43]	16/M	Mandible	Well circumscribed radiolucent circular lesion	Myofibroblastic spindle cells with scattered lymphocytes	IMT	6 months
Biniraj et al / 2014[44]	38/F	Maxillary sinus	MRI – T1 weighed image with contrast showing soft tissue lesion	Fasicles of proliferating spindle cells admixed with inflammatory cells	IMT	NA

Adachi et al., / 2015[45]	42/M	Mandible	MRI – Low 8ntensity on an enhanced T1 image and an enhanced margin of the lesion on a gadolinium enhanced T1 weighed image	Fasicles of spindle shaped cells admixed with dense inflammatory cells	IMT	1 year
Nitesh et al., / 2015[46]	70/M	Retromolar area	NA	NA	IMT	NA
Alkindi et al., / 2016[47]	27/F	Mandible	MRI – Focal area of solid homogenous tissue	Myofibroblastic spindle cell proliferation admixed with chronic inflammatory cell infiltrate	IMT	2 years
Dodhia et al.,/ 2016[48]	10/M	Tongue	NA	Moderately cellular fascicles of spindle cells with myofibroblastic appearance and scattered lymphocytes	IMT	6 months
Tateishi et al., / 2016[49]	11/F	Mandible	CT – Well circumscribed non expansile 3 cm osteolytic lesion	Plump spindle cell proliferation in a storiform pattern and lymphocytes scattered around the spindle cells in a myxoid background	IMT	18 months
Andrade et al / 2017[50]	35/M	Mandible	CBCT – Lytic expansile mass in the anterior and right posterior mandible	Spindle shaped myofibroblast cells and chronic inflammatory cells	IMT	1 year
Kolrepara et al., / 2017[51]	22/M	Mandible	OPG – Irregular radiolucency extending from distal aspect of 38 to posterior border of ramus	Fasicles of elongated spindle cells in a fibrous stroma admixed with intense inflammatory cells infiltrate	IMT	NA
Bajrang P sha et al., / 2017[52]	30/F	Tongue	MRI – Heterogenous hyperintense on T2 weighed image, hypointense on T1 weighed image.	Spindle to oval cells admixed with lymphocytes, plasma cells and eosinophils.	IMT	2 years
Chiara caporalini et al., / 2018[53]	10 mon/F	Tongue	MRI – Hyperintensity on T2 weighed images and STIR sequences after contrast injections.	Spindle shaped cells admixed with plasma cells, lymphocytes, eosinophils	IMT	1 year
Louis monteiro et al., / 2019[54]	16/M	Buccal mucosa	NA	Fusiform cells with histiocyte and lymphoplasmic inflammatory infiltrate	IMT	6 months
Supraj J Shetty., / 2019[55]	29/M	Gingiva	NA	Compact cellular spindle cell proliferation in storiform pattern intermixed with inflammatory infiltrate	IMT	NA

Jose Louis Trevino Gonzalez et al., / 2020[56]	43/M	Tongue	Insignificant	NA	IMT	6 months
Mohammed asifur rahman / 2020[57]	40/F	Mandible	CT – Hypodense area involving molar region	Spindle shaped cells in a fasicular growth pattern admixed with lymphocytes, plasma cells, neutrophils and eosinophils.	IMT	1 year
Vikender S yadav et al., / 2020[58]	50/F	Gingiva	Insignificant	Subepithelial fibroblastic lesion infiltrated with chronic inflammatory cells	IMT	2 years
Eley et al., / 2020[59]	29/M	Buccal alveolus	Insignificant	Stellate and bipolar spindle cells in a myxoid stroma	IMT	6 years
Dhivakar CP et al., / 2021[60]	22/M	Gingiva	NA	Proliferating spindloid fibroblast and myofibroblasts with dense inflammatory infiltrate.	IMT	NA
Nurhayu ab rahman et al., / 2022[61]	70/M 55/M	Tongue Palate	Insignificant	Infiltrative proliferation of spindle cells in a fasicular and storiform pattern. Cellular spindle cells arranged in long fascicles, broad sheets and haphazard pattern.	IMT IMT	2 months 12 weeks
Demet etit et al., / 2022[62]	58/F	Buccal mucosa	MRI – intense homogenous contrast involvement	Spindle shaped myofibroblast like cells intermixed with inflammatory cells.	IMT	3 months
Patil PH et al., / 2022[63]	8/M	Retromolar region	Insignificant	Spindle cells in interlacing fascicles are seen in subepithelial location	IMT	6 months
Goyal et al., / 2022[64]	27/F	Gingiva	Insignificant	Highly cellular stroma with plasma cells and subepithelial spindle cells arranged in fasicles	IMT	2 years
Limi Diaeo et al., / 2023[65]	47/M	Submandibular gland	CT – isodense nodular shadow in the submandibular gland	Spindle and oval shaped cells were arranged in sheets and intersecting fascicles admixed with inflammatory cells	IMT	13 months

Present Case	9/F	Buccal mucosa	Ultrasonography – Well defined heteroechoic solid lesion	Irregularly arranged fascicles of spindle cells and inflammatory cells	IMT	1 year

NA – Not available

Table 2 – Differential diagnosis of inflammatory myofibroblastic tumour with key differentiating feature

Nodular fasciitis	Common in extremities. Feathery appearance with C-shaped fascicles and mucin rich stroma with less prominent inflammatory infiltrate. ALK negative.
Solitary fibrous tumour	Areas resembling hemangiopericytoma with strong immunopositivity for CD34.
Benign fibrous histiocytoma	Typical storiform pattern
Calcifying fibrous tumour	Hypocellular benign neoplasm. Scattered dystrophic calcification is seen.
Desmoid type fibromatois	Spindle cells with interspersed collagen with minimal to no inflammation and negative for ALK.
Myofibroma	Biphasic growth pattern with hemangiopericytoma like blood vessels
Fibrosarcoma	Collagenous area with herring bone pattern with less prominent inflammatory cells.
Follicular dendritic cell tumour	Dendritic spindle cells admixed with inflammatory infiltrate
Granulocytic sarcoma	Dense subepithelial infiltrate of pleomorphic cells
Inflammatory leiomyosarcoma	More organized fascicular growth with cigar shaped nuclei.
IgG4 related sclerosing disease	The IgG4+/IgG ratio is significantly higher than in IMT and is negative for ALK.
Hodgkins lymphoma	Shows positivity for CD15 and CD30 and negativity for ALK 1.
Inflammatory fibroid polyp	Myxoid stroma with lymphocytes and eosinophils.
Gastrointestinal stromal tumour	Inflammation is rich in lymphocytes, with rare eosinophils or plasma cells. DOG1 and CD117 are positive while ALK is negative.

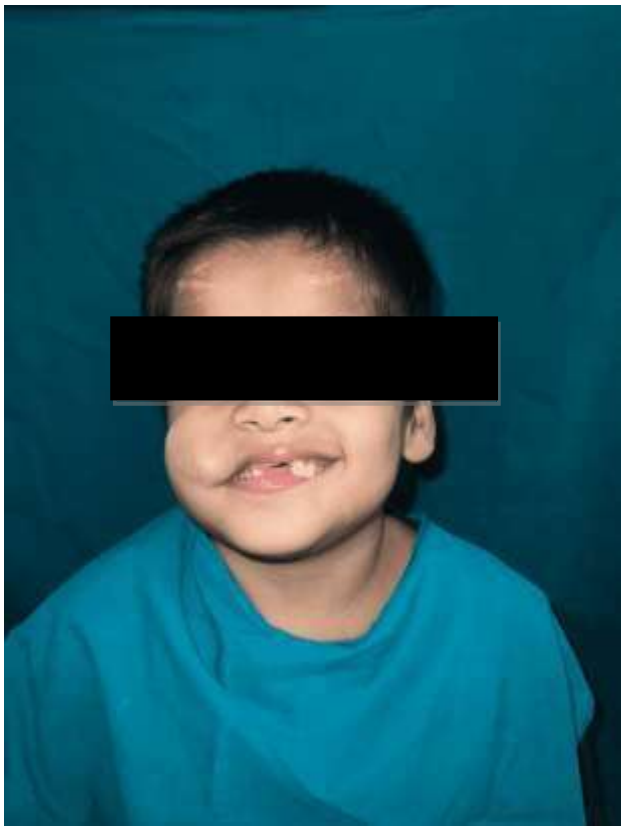

Fig 1. Gross image showing swelling and facial asymmetry

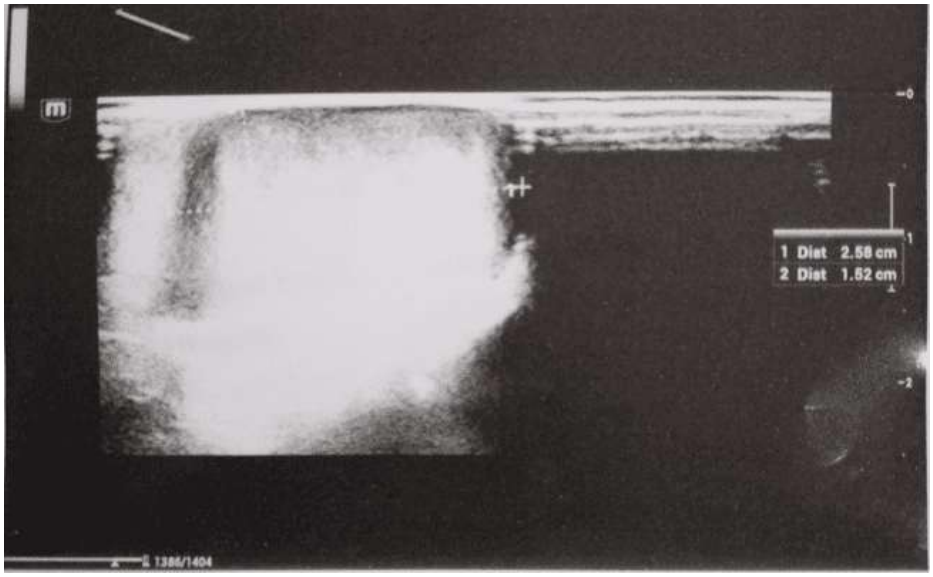

Fig 2. Ultrasonographic image showing heteroechoic solid lesion

Fig 3. Gross image of the lesion

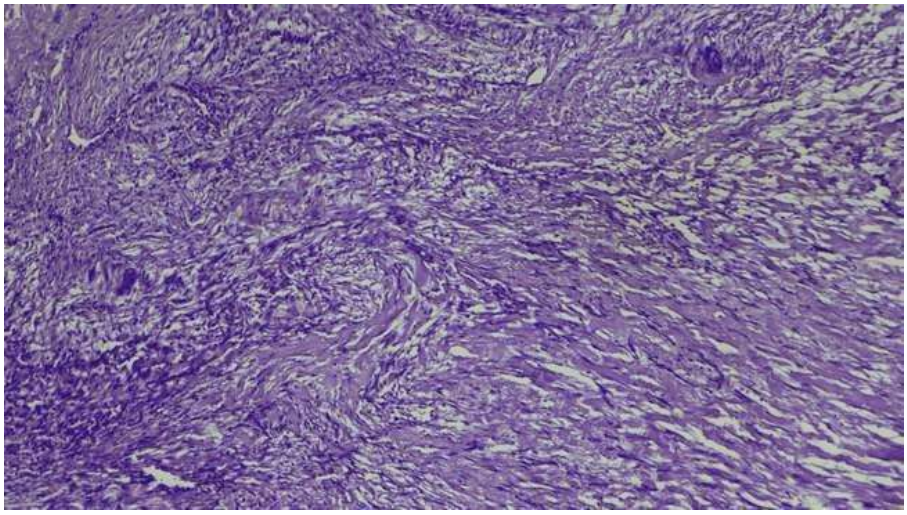

Fig 4. H&E staining shows irregular fascicles of spindle cells and inflammatory cells at 10x.

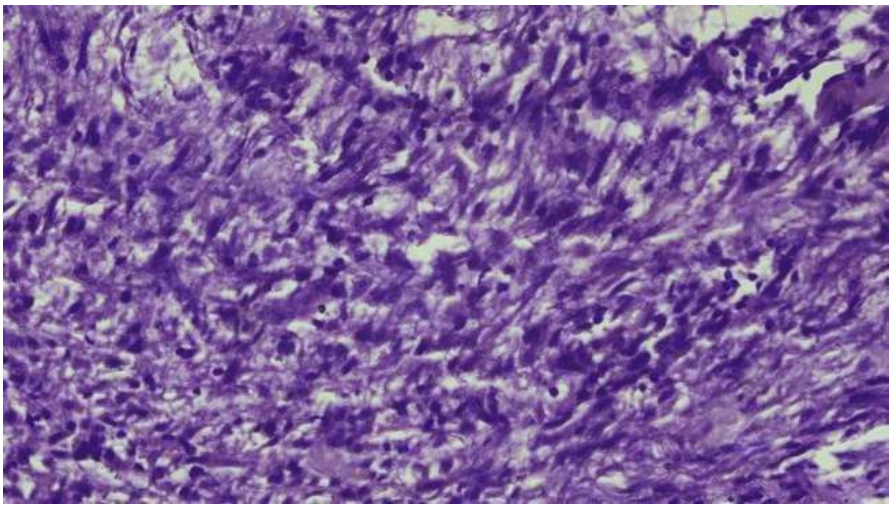

Fig 5. H&E image showing fascicles of spindle cells and inflammatory cells at 40x.