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6 Nodular Fasciitis and Myxolipoma of the Larynx

7 *A rare case report with brief literature review*

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16 **Abstract**

17 Nodular fasciitis (NF) is a peculiar, rapid-growing soft tissue lesion, typically appearing in
18 subcutaneous tissue. 20% of NF occur in the head and neck region, where they can involve any
19 anatomic site. Laryngeal involvement, however, is quite rare. On the contrary, Lipoma is
20 recognized as a slow growing, benign mesenchymal tumour. Myxolipoma is a rare variant which
21 has a prominent myxoid background. Laryngeal lipoma is infrequent, accounting for only 0.6%
22 of all benign laryngeal lesions. Here, we report a unique case of adult laryngeal nodular fasciitis
23 coexisting with myxolipoma in a 61-year-old male patient, describing their clinical and
24 histopathological features, the strategies used to treat such conditions along with a brief review
25 of the literature. The purpose is to broaden the differential diagnosis of rapid-growing laryngeal
26 masses that cause airway obstruction and to stress the significance of integrative interdisciplinary
27 collaboration on reaching an accurate diagnosis, thereby allowing proper management for benign
28 pathologies and avoiding any futile aggressive treatment.

29 **Keywords:** Nodular fasciitis, Larynx, Stridor, Myxolipoma.

31 **Introduction**

32 Nodular fasciitis, a subtype of benign mesenchymal spindle cell tumor-like lesions, is a
33 noncancerous, reactive fibrous proliferation which makes up almost 11% of all benign soft
34 tissue tumours.¹

35
36 Although Konwaler et al. first described this condition as ‘pseudosarcomatous fasciitis’ in
37 1955, it was not until 1961 that Shuman used the term ‘nodular fasciitis’ for the very first time,
38 and since then it has been broadly adopted by authors.²

39
40 It arises mostly in the extremities and trunk,³ with the larynx being a very rare location for
41 NF, so diagnosis of NF is challenging in this region. In most cases, the patient presents with a
42 painless, rapidly expanding subcutaneous mass. However, unlike other locations, laryngeal NF
43 may cause life-threatening symptoms by obstructing the respiratory tract.

44
45 In contrast, lipoma is a common slow developing benign mesenchymal tumour, with roughly
46 15% of lipomas being found in the head and neck. Due to the presence of scant amounts of
47 adipose tissue in the larynx, laryngeal occurrence is infrequent.⁴ Myxolipoma is a rare variant
48 which has prominent myxoid areas.

50 **Case Report**

51 A 61-year-old male, who is a heavy smoker, presented to the ENT outpatient clinic in King
52 Abdullah University Hospital, October 2020, complaining of a change in his voice, which had
53 started few months earlier, associated with dysphagia, intermittent dyspnoea and choking. His
54 surgical history was unremarkable except for laparoscopic cholecystectomy 1 year ago. There
55 was no history of trauma. Head and neck examination was unremarkable. A large right
56 supraglottic mass was identified by flexible nasolaryngoscopy. Radiological findings were
57 suggestive of a locally advanced laryngeal cancer (**Figure 1**). Biopsy was taken by suspension
58 microlaryngoscopy for precise diagnosis and results were suggesting a benign/borderline spindle
59 cell neoplasm.

60
61 Since it has a benign nature, lesion was debulked trans-orally by laser and histologic diagnosis
62 was in favour of laryngeal nodular fasciitis. Shortly after surgery, the mass rapidly enlarged,

63 tracheostomy was performed and he underwent complete mass excision with right partial
64 laryngectomy through an open surgical approach (**Figure 2**). Repeated pathological examination
65 of microscopic sections and immunohistochemistry study revealed same findings in accord of the
66 aforementioned diagnosis of laryngeal nodular fasciitis, along with incidental myxolipoma
67 (**Figure 3&4**). Postoperatively, the patient recovered uneventfully with no evidence of
68 recurrence. He is currently much better and under regular follow up. Consent was obtained from
69 the patient to publish this case report.

70

71 **Discussion**

72 **Clinical features**

73 Nodular fasciitis is a rare, but distinct, benign self limited mesenchymal neoplasm of
74 fibroblastic/ myofibroblastic derivation that resembles soft tissue sarcoma. The etiology is still
75 not fully understood. Although not documented in the vast majority of patients, many believed
76 that traumatic insult is the trigger for this reactive inflammatory response on most occasions.³
77 While occurring at all ages, this condition is most often diagnosed in adults 20 to 40 years of age,
78 with no gender predilection.⁵

79

80 Clinically, when appearing in the larynx, the main presenting symptoms are related to the degree
81 of laryngeal obstruction. It can include hoarseness of voice, foreign body sensation, dysphagia,
82 as well as dyspnoea and stridor that may require urgent medical attention. Despite most of the
83 lesions being less than 2 cm in greatest dimension at the time of diagnosis, the size may vary
84 from 0.5 to 10 cm.⁶

85

86 Equivalently, lipoma is a benign tumor consisting of adipose tissue. It is considered the most
87 common mesenchymal tumor, constituting 16% of soft tissue tumors.⁶ Lipoma of larynx can be
88 subdivided according to the site of origin into Intrinsic (Endolaryngeal) type, or more commonly
89 Extrinsic type. It has a male predominance and occurs over a wide age range (mean age 40 years)
90 with a supraglottic predisposition.⁴

91

92 The etiology of laryngeal lipomas is unclear. Unlike other locations, laryngeal lipomas may
93 occasionally cause fatal airway obstruction with dyspnea and dysphonia being the most frequent

94 presenting symptoms. Grossly, it can be sessile or pedunculated, usually appearing as a smooth,
95 well-encapsulated mass. The size typically ranges from 1 to 3 cm, but sometimes it may exceed
96 10 cm.⁷

97

98 **Radiological findings**

99 On CT scan most of NF lesions appear as a well-defined homogeneous mass with low or
100 isodensity and show moderate to strong enhancement. On MRI lesions will exhibit hypointense
101 or isointense signals on T1-weighted sequences and show heterogeneous intermediate-to-high
102 signal on T2-weighted sequences.^{8,9}

103

104 For lipomas, imaging serves a vital role in diagnosis. On CT scan it mostly simulates fatty tissue
105 characteristics, thus lipomatous lesions appear as a homogenous mass with low attenuation. At
106 MRI, lipoma has predominantly low signal intensity on T1-weighted images and markedly high
107 signal intensity on T2-weighted images.¹⁰

108

109 **Histopathology findings**

110 Lesions of NF are often composed of undulating short, intersecting fascicles of haphazardly
111 arranged plump, immature fibroblasts and myofibroblasts in a loose myxoid and/or fibrous
112 stroma which resembles the feathery or tissue culture-like appearance.

113

114 The cells have uniform, elongated non-pleomorphic nuclei with pale, fine chromatin and small
115 but prominent nucleoli. Some typical mitotic activity is commonly seen. Numerous extravasated
116 red blood cells, scattered lymphocytes, chronic inflammatory cells and multinucleated osteoclast-
117 like giant cells are also present within the background.¹¹

118

119 Likewise, lipomas have some distinctive histological features, being generally composed of
120 mature adipocytes bound by thin fibrous capsules. Myxolipoma demonstrates similar features,
121 but with abundant extracellular mucoid matrix.¹²

122

123 **Immunohistochemistry findings**

124 Myofibroblasts often stain for Smooth Muscle Actin, muscle-specific actin and Vimentin.
125 Meanwhile, none of lesional cells express S100 protein, β -catenin, CD34, keratin, caldesmon and
126 desmin. The proliferation index with Ki67 can be high in reactive lesions such as NF.¹³ It should
127 be noted that Immunohistochemistry is not of much help in diagnosing myxolipoma.

129 **Molecular and cytogenetic findings**

130 In regard to the post-genomic era and cytogenetic tests, few studies established the molecular
131 and cytogenetic abnormalities and proved the neoplastic nature of NF. For example, Erikson-
132 Johnson MR et al. described the USP6 rearrangement with the formation of the fusion gene
133 MYH9-USP6 which is commonly observed in the lesions, and they referred to nodular fasciitis
134 as ‘transient neoplasia’ in tribute to its self-limiting nature.¹⁴

136 **Treatment and prognosis**

137 Given the paucity of cases of NF involving the larynx, their natural course is not fully
138 understood. However, assuming that their behavior would be as with NF of other anatomic sites,
139 as this lesion had neither a high local recurrence propensity nor metastatic potential, adequate
140 surgical excision of the lesion with negative margins could be sufficient.

142 Yet, due to anatomic factors and critical structures, laryngeal NF lesions can be unamenable to
143 simple complete laryngoscopic local excision. Hence, Partial laryngectomy, whether endoscopic
144 or open (based on the lesion, surgeon’s skills and patient factors) could be an appropriate choice
145 in favour of vocal function preservation. Total laryngectomy should be used in selected cases
146 with advanced diseases or reserved as salvage surgery.

148 Other controversial conservative methods like intralesional corticosteroid injection can be
149 considered when there are no substantial symptoms and spontaneous regression is expected.
150 Surgical excision is recommended in the case of laryngeal lipoma. Depending on the size,
151 endoscopic approach is preferred in small lipomas, whereas open surgical approach must be used
152 if the lesions dimension is beyond 2 cm. As for benign lesions, the prognosis for laryngeal
153 lipoma is very good. Recurrence is rare and is mostly due to hidden malignancy or inadequate
154 excision. Because of this, long-term follow-up is recommended.¹⁵

155

156 **Conclusion**

157 Nodular fasciitis of the larynx can mimic malignant tumors, thus reaching an exact diagnosis is
158 very challenging. Although it has a favorable prognosis as compared to other aggressive
159 laryngeal lesions, simple lesional resection in this unique location might be difficult to obtain
160 and laryngectomy is inevitable. Laryngeal myxolipoma is rare but must be considered in the
161 differential diagnosis of laryngeal masses. To the best of our knowledge, only four cases of
162 laryngeal myxolipomas, one of them being juvenile, have been reported in the English literature.

163

164 **Statement of Ethics:**

165 The patient has given his written informed consent to publish his case (including publication
166 of images).

167

168 **Author's Contribution:**

169 AO and RAA performed the literature review. All authors contributed to the drafting of the
170 manuscript. All authors approved the final version of the manuscript.

171

172 **Referances**

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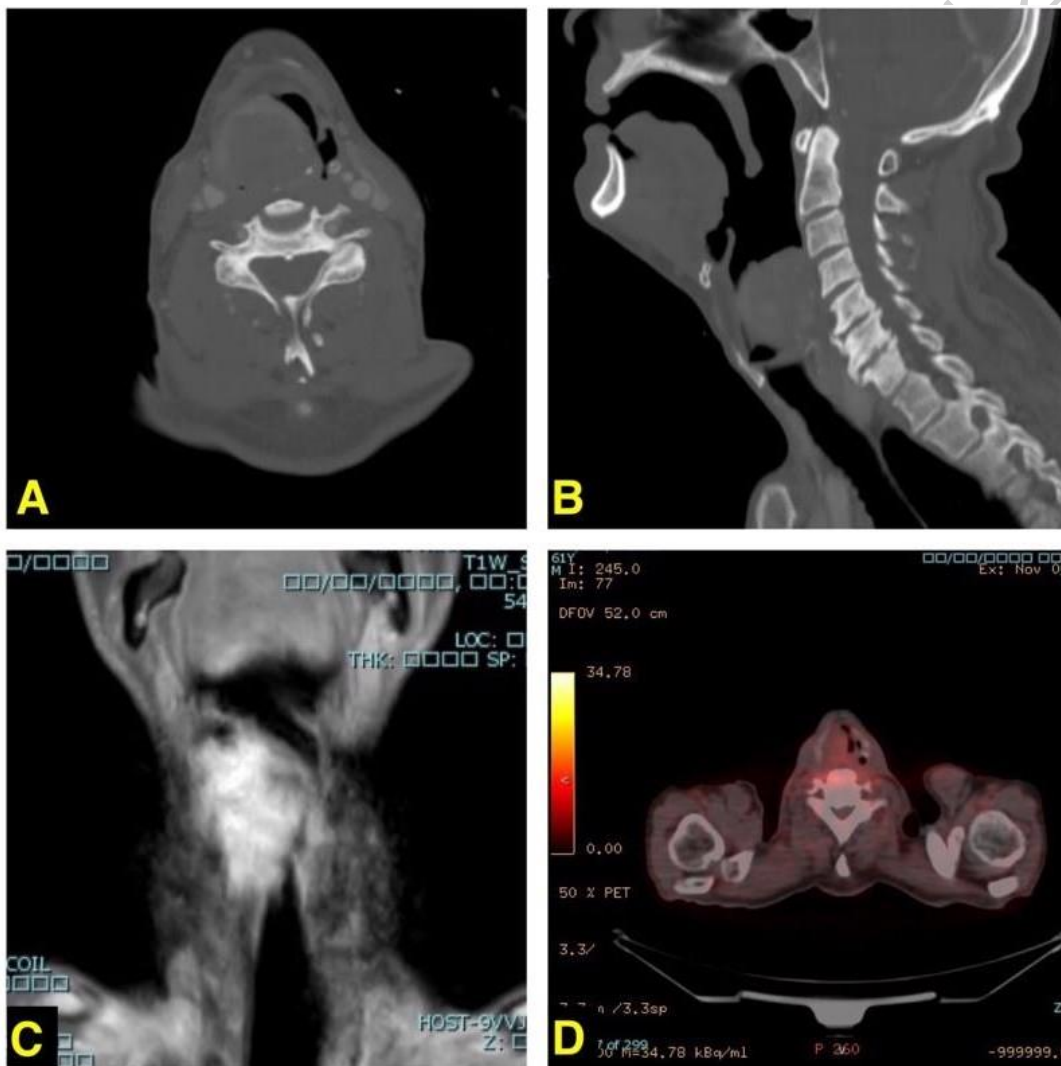
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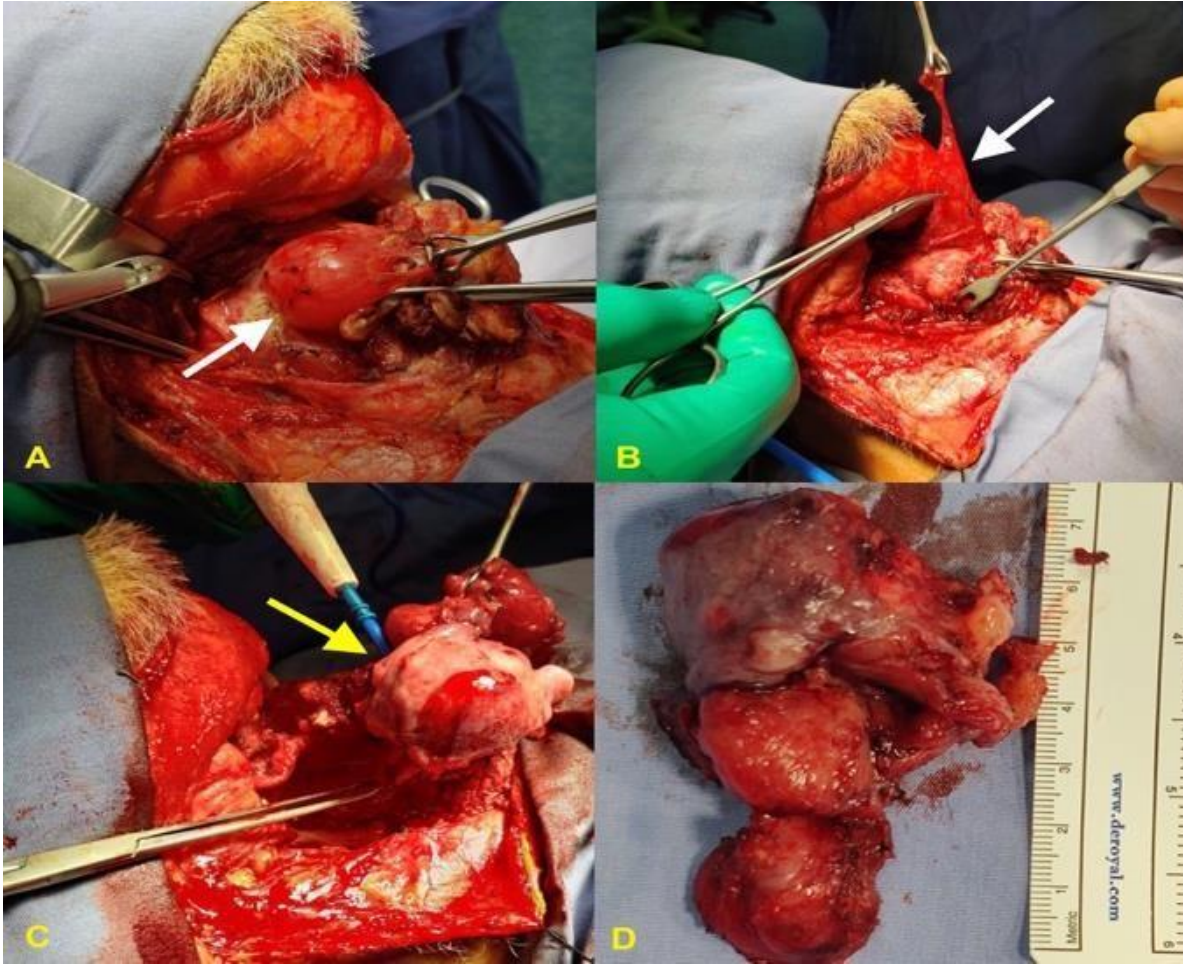
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222 **Figure 1:** (A) Axial, (B) sagittal CT scan showed a heterogenous laryngeal mass with thick-
223 enhancing rim, measuring about 5.1 x 3.8 x 3.7 c.m starts at the level of the hyoid bone and
224 extends caudally reaching the right cricoid cartilage. (C) Coronal view on T1W MRI showed the
225 same mass causing compression effect on the adjacent vessels with significant stenosis on supra-

226 glottic region and minimal invasion of the right thyroid cartilage. **(D)** PET scan revealed a
227 hypermetabolic laryngeal mass.

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229

230 **Figure 2:** **(A+B)** Intraoperatively, cystic lesion with extra-laryngeal extension incidentally
231 found, mimicking laryngocele (white arrow). **(C)** Another smooth, well circumscribed, firm
232 mass which seemed to involve the right thyroid cartilage and reach to the false vocal cords was
233 identified (yellow arrow). **(D)** Excised specimen which includes part of larynx with cystic
234 structure attached to a well-defined mass.

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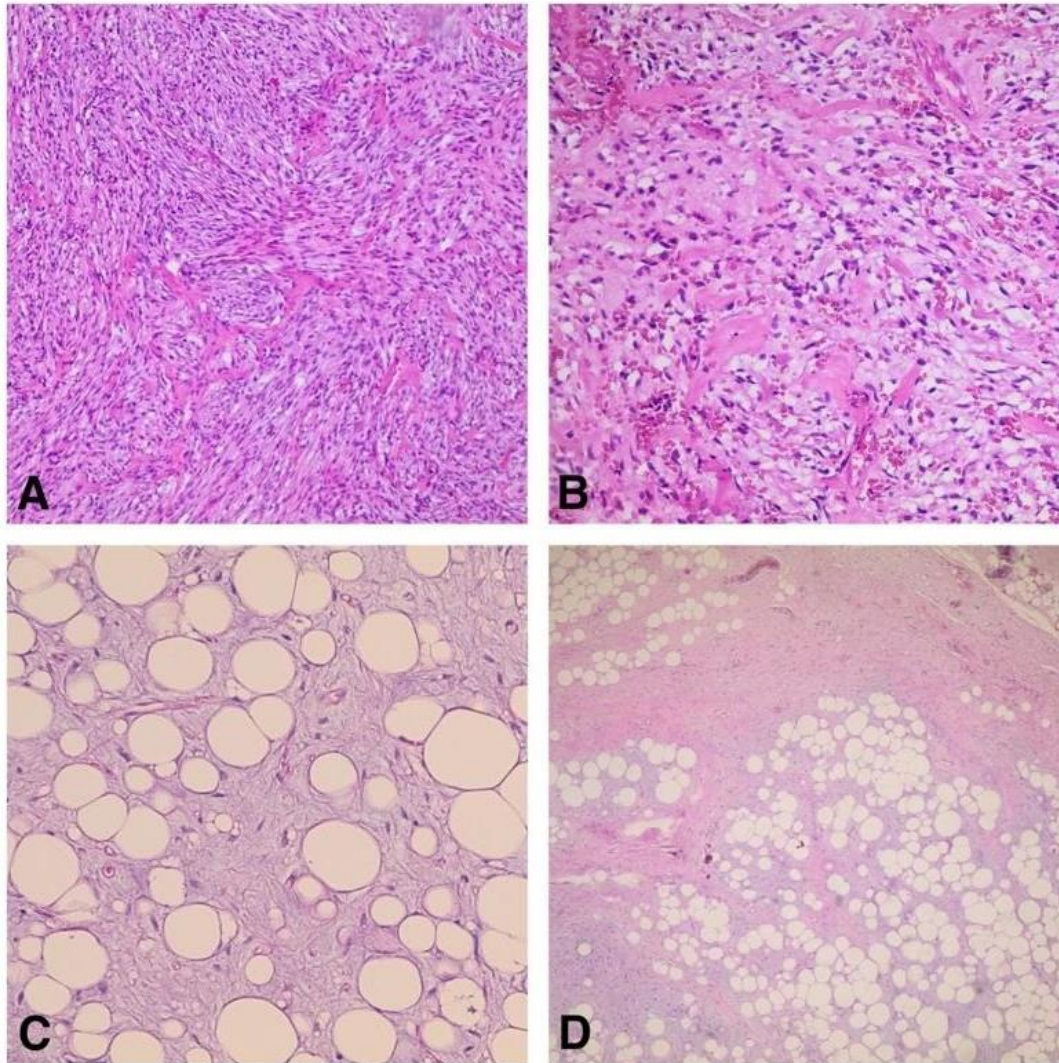
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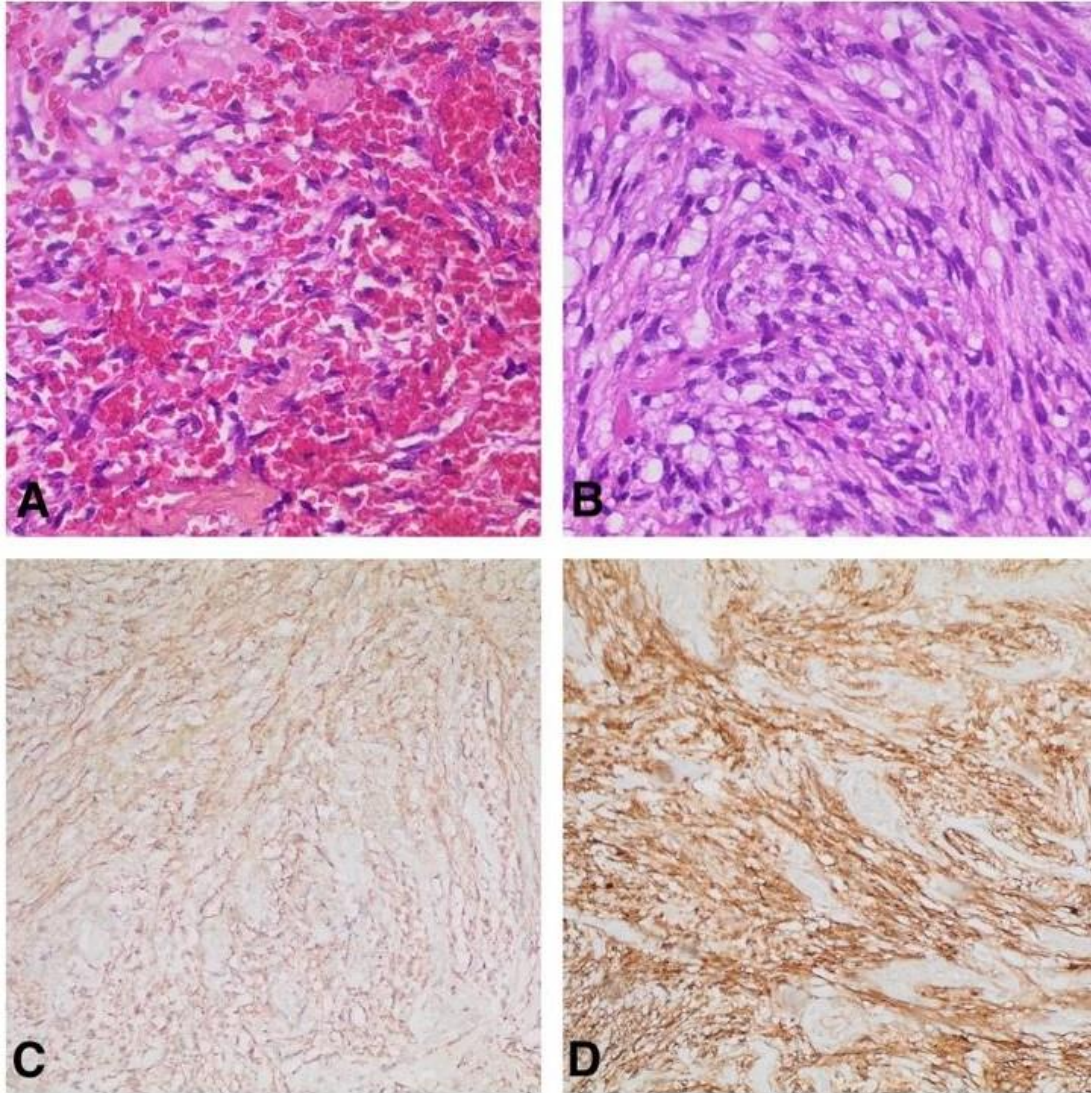
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244 **Figure 3:** H&E image shows (A) Spindled and plump cells exhibiting fascicular arrangement in
245 myxofibrotic background. Lesional cells demonstrate storiform architecture. (B) Benign looking,
246 uniform, stellate fibroblasts resembling tissue culture-like appearance. (C+D) Myxolipoma
247 consists of mature-appearing adipocytes with prominent myxoid areas. No evidence of atypia,
248 plexiform vascular network or lipoblasts. (A: 20 x / B,C: 40x / D: 10 x magnification).

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256 **Figure 4:** H&E image; 40 x magnification, shows (A) Numerous extravasated red blood cells
257 and scattered chronic inflammatory cells. (B) Lesional cells of nodular fasciitis typically have
258 ovoid nuclei and prominent nucleoli. Immunohistochemistry study revealed diffuse expression of
259 SMA Immunohistochemical marker in the lesional cells (C) and CD10 positive immunostaining
260 of nodular fasciitis (D).