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Extranuchal-type Fibroma Mimicking Low Grade Sarcoma: A Case Report and Review of the Literature

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Abstract Nuchal-type fibroma (NTF) is a rare fibrous growth that developing in the nuchal region, in some instances it occurs at the extranuchal sites. Herein, this study reported a case of NTF on extranuchal site in an Iranian girl in Western Iran. The report included a 14-year-old girl with a chief complaint of right hemithorax suprascapular soft tissue mass since 6 years ago and tenderness since last month that physical examination showed firm, tender, non-erythematous mass measuring 5x10 cm with adhesion to surrounding tissue was noted on the right hemithorax over the scapula. Frozen section diagnosis showed spindle cell tumor compatible with low grade sarcoma, but the pathologist reported an extranuchal-type fibroma in permanent. In conclusion, NTF occurs more frequently in middle-aged adults and is more common in men. The prevalence of nuchal region is more than extranuchal region in NTF patients. The NTF is associated with trauma more than Gardner's syndrome. Also, the pathologist and surgeon must be aware of this entity, especially at extranuchal sites, not to be misinterpreted as sarcoma.

Keywords: *extranuchal-type fibroma, low grade sarcoma, case report*

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1. Introduction

Nuchal-type fibroma (NTF) is a distinct subcutaneous and the dermal fibrous tissue proliferation [1] and was first described by Enzinger and Weiss in 1988 [2] that is a rare fibrous growth occurring predominantly in the interscapular and paraspinous regions [3]. Although NTF typically develops in the nuchal region, in some instances it occurs at the extranuchal sites [4]. This mass is characterized by hypocellular bundles of collagen with entrapped adipocytes and increased numbers of small nerves [5]. It is often associated with trauma or infection and sometimes accompanied by the feeling of discomfort or pain [6] and also can be associated with diabetes mellitus, scleredema or Gardner's syndrome [7]. The aim of this study was to report a case of NTF on extranuchal site in an Iranian girl in Western Iran.

2. Case Report

A 14-year-old girl was admitted to The Department of Surgery on 6th August, 2016 with a chief complaint of right hemithorax suprascapular soft tissue mass since 6 years ago and tenderness since last month. There was no significant past medical history or drug history. On physical examination; firm, tender, non-erythematous mass measuring 5x10 cm with adhesion to surrounding tissue was noted on the right hemithorax over the scapula. Otherwise, physical examination was unremarkable. The lab data, such as complete blood count, thyroid function tests, biochemistry and coagulative function analysis were normal. The patient underwent surgery on 7th August and the first specimen was sent for frozen section diagnosis with clinical impression of sarcoma. Grossly the specimen was consisted of white homogenous mass measuring 13x8.5x3.5 cm (Figure 1). Frozen section diagnosis showed spindle cell tumor compatible with low grade sarcoma and recommendation of waiting for permanent diagnosis and immunohistochemistry (IHC). The patient was discharged on 9th August with good condition. The pathology report on 28th August was compatible with extranuchal-type fibroma (Figure 2). The IHC staining of CD34 was positive and SMA (smooth muscle actin) was negative in favor of diagnosis (Figure 3).

3. Discussion

This case reported NTF of right scapula with 6-year duration of mass without a significant past medical history in a 14-year-old girl that physical examination showed firm, tender, non-erythematous mass measuring 5x10 cm. Table 1 and Table 2 show the characteristics of case reports and case series of the NTF, respectively. This tumor occurs more frequently in middle-aged adults and is more common in men [8]. The mean age of the patients is 39.8 years (range, 3-74 years). Out of 83 patients, 69 (83.1%) are men and 14 (16.9%) women. The duration range of disease is ~1 to 30 years. Also, NTF typically

develops in the nuchal region, but in approximately one-third of the cases, it occurs at extranuchal sites [4] and the most common extranuchal sites of NTF are the back or scapular region, shoulder, and face [9]. Out of 78 lesions in 75 patients; 44 lesions (56.4%) were in nuchal region and 31 lesions (43.6%) extranuchal region that the most common extranuchal region was shoulder. The size range of tumor is 1 to 30 cm. Although the pathogenesis of NTF remains unknown, it has been associated with trauma [7,8,10,11,12] and rarely Gardner's syndrome [1,7,8]. Out of 8 case reports and a case series with 7 patients, 3 (20%) patients had Gardner's syndrome and out of 9 case reports and a case series with 7 patients, 9 (56.2%) patients had trauma. Also, Michal et al. [8] reported that 3/52 (5.8%) lesions and 47/52(91.3%) had Gardner's syndrome and trauma, respectively.

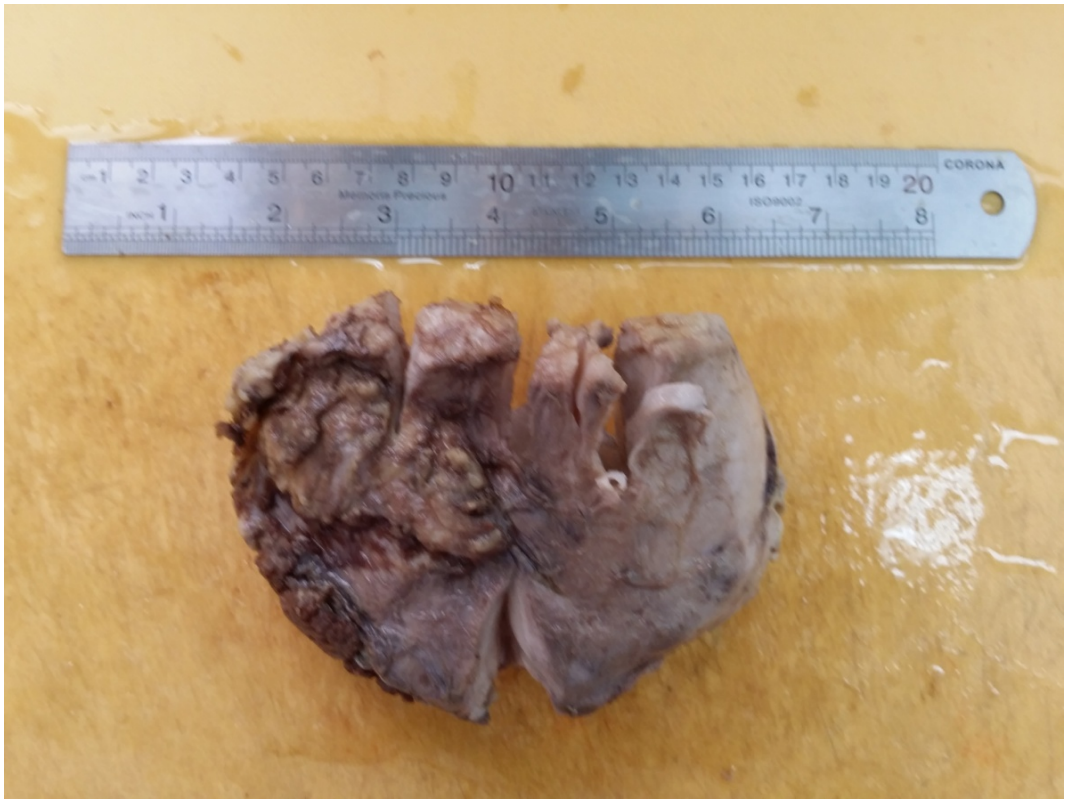

Figure 1. Gross specimen, extranuchal-type fibroma mimicking low grade sarcoma

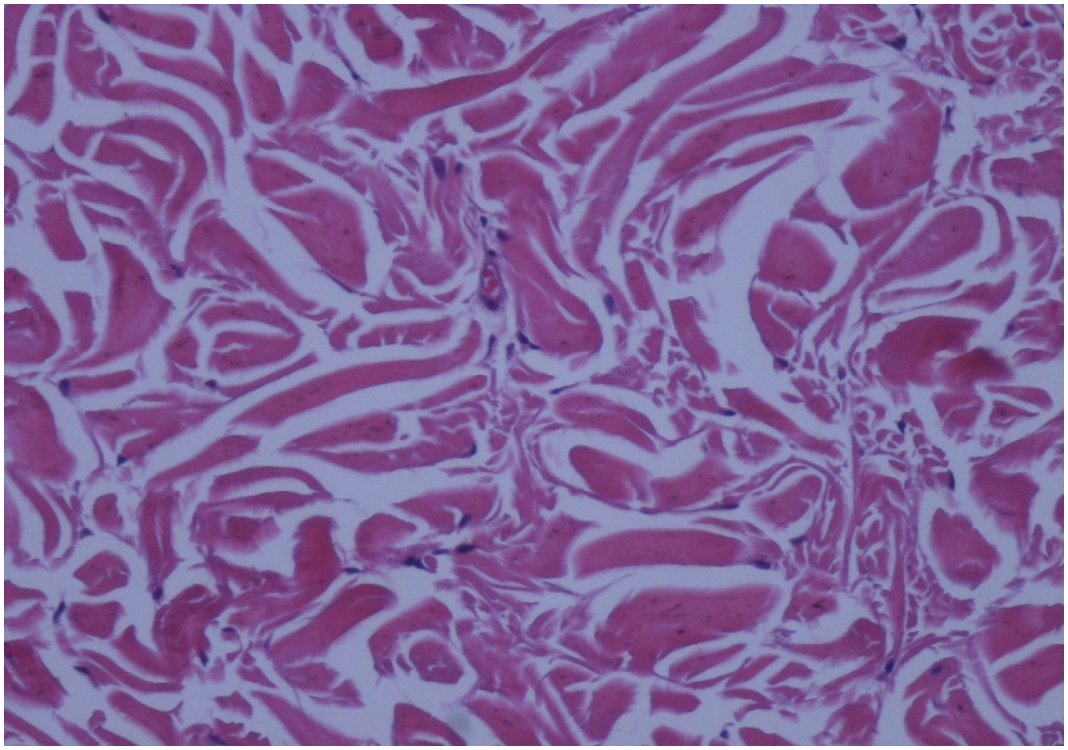

Figure 2. Hypocellular dense collagen bundles with scattered fibroblasts (Hematoxylin & Eosin, x200 magnification)

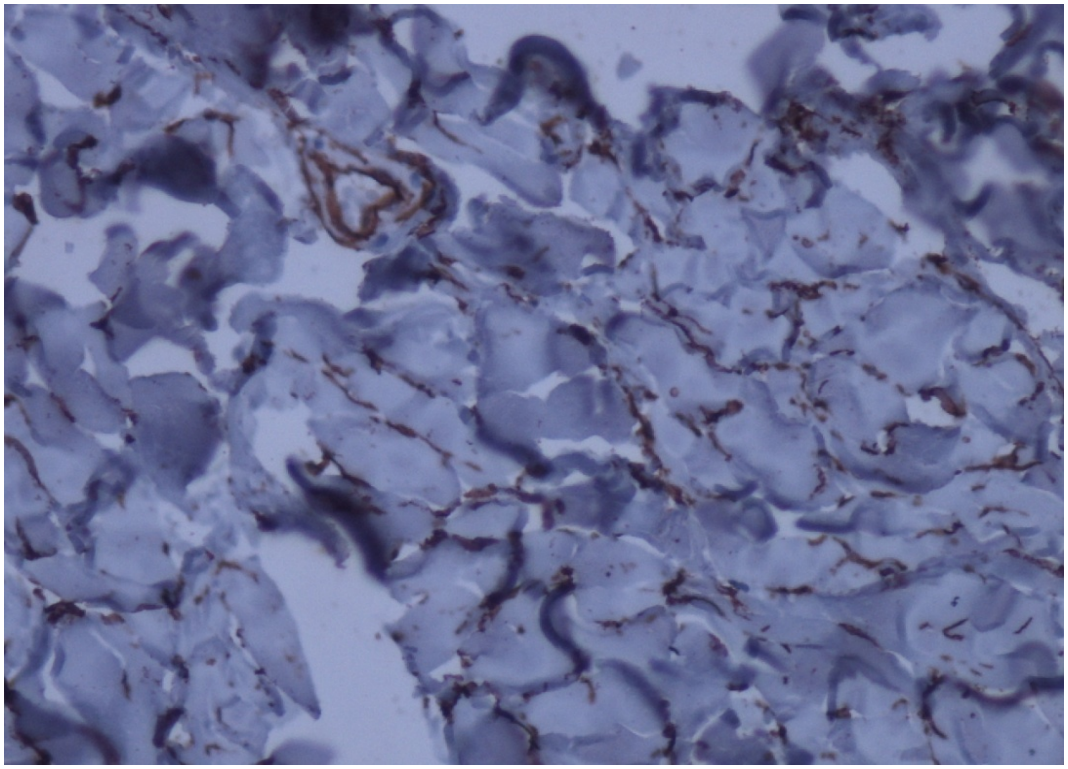

Figure 3. CD34 positivity in spindle cells (Immunohistochemistry, x200 magnification)

Table 1. The characteristics of case reports of nuchal-type fibroma

Reference	Age	Sex	Duration, years	Tumor location	Tumor size, cm	Gardner's syndrome/Trauma
Diwan et al. [1]	13	M	?	Right upper back	?	Yes/?
Diwan et al. [1]	60	M	?	Upper chest	?	Yes/?
Lee et al. [3]	45	F	2	Right buttock	3.5x2.6x2.0	?/No
Kim et al. [4]	32	M	6	Scalp	Several	?/?
Gong et al. [5]	48	M	~30	Right shoulder	20x10x5	No/No
Abe et al. [6]	52	M	30	Posterior neck	3.8x3	?/No
Linos et al. [7]	54	M	2	Left buttock	4.8	Yes/Yes
Sraj et al. [10]	20	M	~5	Right ankle	6.0x5.0x1.8	?/Yes
Karonidis et al. [11]	38	M	Several years	Midline posterior neck	3.5x1	No/No
Banney et al. [12]	53	M	30	Midline posterior neck	?	?/?
Diwan and Horenstein [13]	43	M	?	Left back and neck	5 and 3	No/?
Tsunemi et al. [14]	66	M	3	Posterior neck	3x2	?/?
Alsaleh and Amanguno [15]	39	F	10	Right posterior neck	30x15	?/?
Hameed et al. [16]	15	M	~1	Left shoulder	15x10	No/No
Lee et al. [17]	35	F	17	Posterior neck	3.5x5	?/No
Yokoyama et al. (18)	62	M	?	Right posterior neck	2	?/?
This case	14	F	6	Right scapula	13x8.5x3.5	No/No

Table 2. The characteristics of case series of nuchal-type fibroma

Reference	Cases	Age, years	Male/Female	Size, cm	Tumor location, N	Mean duration /range, years	Gardner's syndrome/Trauma
Michal et al. [8]	50 (52 lesions)	Mean:40 Range: 3-74	41/9	Mean:3.2 Range:1-6	Nuchal:36; Extranuchal:16 (back or scapular:6; shoulder:3; face:3; left forearm:1; anterior neck:1; knee:1; truncal:1)	Long duration	3 lesions/47 lesions
Lee et al. [19]	7	Mean:26.8 Range: 21-32	7/0	Mean:9.1 Range:7.5-12	Right shoulder:3; Left shoulder:3; Right and Left shoulder:1	8.1/4-12	No/Yes
Balachandran et al. [20]	9	Median:43 Range: 19-53	8/1	Median:3.5 Range:2.5-8	?	?/?	?/?

The role of trauma in the etiology of nuchal-type fibroma is unknown [12]. Consistency, mobility and tenderness in physical examination of this tumor is different including firm [4,7,13,11], mobile, and mildly tender [13], solid, immobile [10], nontender subcutaneous mass [3,11] and elastic, hard nodule [14] that this case reported firm, tender, non-erythematous mass.

Microscopic examination of the mass is composed of thick bundles of dense collagen interspersed with hypocellular [3,4,10,11,14,15], bland spindle cells with small uniform nuclei [10], and is mainly poorly circumscribed [4,14]. Also, gross specimen has different colors and circumscriptions such as white and pearly appearing than the deeper off-white tissue with some yellow specks/ poorly circumscribed [13], tan to-white cut surface/ irregular margins, in both the dermis and subcutis [7], and tan to grayish lobulated mass [15] that may be mistaken for sarcoma. This case reported white homogenous mass measuring 13x8.5x3.5 cm that due to the large size, color and poor circumscription was mistaken for sarcoma in frozen section interpretation.

4. Conclusions

NTF occurs more frequently in middle-aged adults and is more common in males (male/female ratio:5/1). The prevalence of nuchal region is more than extranuchal region in NTF patients. The NTF is associated with trauma more than Gardner's syndrome (about 3-fold more). Also, the pathologist and surgeon must be aware of this entity, especially at extranuchal sites, not to be misinterpreted as sarcoma.

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