MERIT RESEARCH JOURNALS
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Merit Research Journal of Medicine and Medical Sciences (ISSN: 2354-323X) Vol. 4(12) pp. xxx-xxx, December, 2016
Available online http://www.meritresearchjournals.org/mms/index.htm

Review

Periorbital Nodular Fasciitis in an Adult Male: The First Reported Case in the Middle East with Review of the Literature

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Abstract

Nodular fasciitis (NF) is a benign, fibroblastic proliferation, which can raise suspicion for malignancy and is relatively rare in the ocular region. We report the first case of periorbital NF in the Middle East with review of the literature. Our case of NF is described clinically and histopathologically in detail. An extensive review of all reported cases of ocular/periocular NF in the English-written literature is made with summary of 39 cases including ours taking into consideration the location of the tumor, age and gender. Our 35-year-old male presented with a recurrent tumor in the periocular region. Diagnosis of NF was made by careful histopathological examination of the excisional biopsy and by immunohistochemical staining. We have summarized the demographics and tumor location of 38 cases previously reported and added our case information. Demographically, NF occurs within a wide range of age (8months to 81years) with a mean age of about 28 years. Half of the cases are in the age group of 30 years or younger (median=30 years). Slight female predominance is observed (F:M =7:6). The right side is more commonly involved (in 21/37). Periocular/orbital involvement is more commonly found (67%).

Keywords: Nodular Fasciitis, Ocular, Periocular

INTRODUCTION

Nodular fasciitis (NF) is an idiopathic, benign, fibroblastic proliferation commonly found in the subcutaneous tissue or superficial fascia of the extremities. These tumors can raise suspicion for malignancy both clinically and histopathologically (Enzinger and Weiss, 1995). The involvement of NF in the orbital region is relatively rare since the initial report by Font and Zimmerman in 1966 (Font and Zimmerman, 1966). We report the first case of nodular fasciitis involving the periorbital region of an adult male in the Middle East.

The Case

A 35-year-old male was referred to our institute for a second opinion. He presented with a slowly progressive left lateral sub-brow lump. The growth was initially noted after trauma to the sub-brow area 3 months prior to his complaint. The patient underwent surgical excisional biopsy under local anesthesia 2 months earlier in an outside center. Later, the patient has noticed recurrence of the same lesion one week after surgery. He denied any history of pain or decrease in vision. There was no significant past medical or ocular history. External examination demonstrated freely mobile non-tender mass measuring 10 X 8 mm over the temporal aspect of the superior orbital rim of the left eye (Figure 1a). His vision was unaffected with no sign of infection and no palpable regional lymph nodes. The rest of his ocular examination was unremarkable in both eyes. Computerized

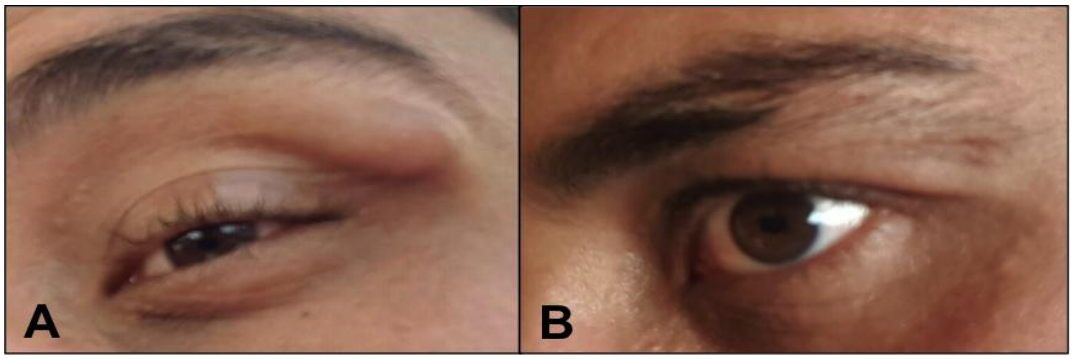

Figure 1a. The clinical appearance of a 10 X 8 mm freely mobile non-tender swelling over the temporal aspect of the superior orbital rim of the left eye.
Figure 1b. A photograph of the same patient six months post-surgery showing good tissue healing and no sign of recurrence.

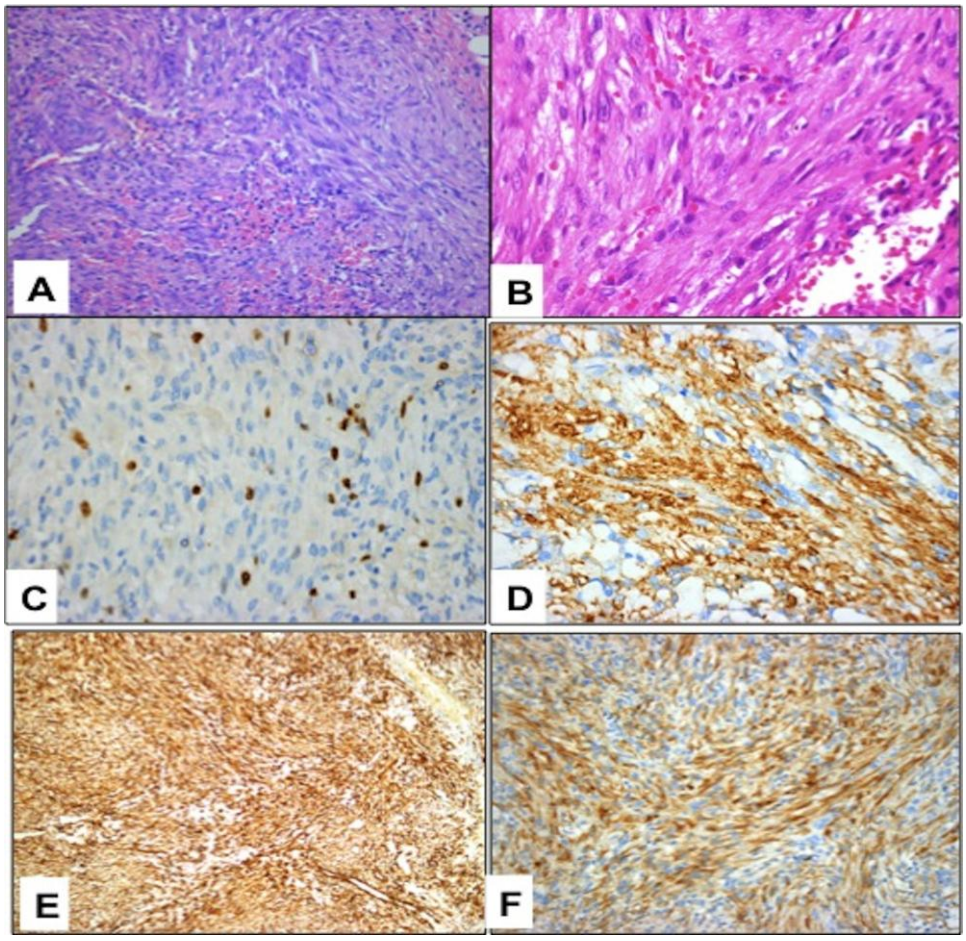

Figure 2a. Histopathology of the excised tissue showing the interlacing bundles of spindle-shaped cells (Original magnification X200 Hematoxylin and Eosin).
Figure 2b. Higher power of the spindle-shaped cells compatible with immature fibroblasts (Original magnification X400 Hematoxylin and Eosin).
Figure2c. The proliferation index of the proliferating cells, which was estimated to be 5 percent (Original magnification X400 Ki67).
Figure 2d. The spindle-shaped cells showing positive staining with slight “Tram track” pattern (Original magnification X400 SMA).
Figure 2e. NF positive cells with Vimentin (Original magnification X 100 Vimentin)**.**
Figure 2f. The spindle-shaped cells showing positive staining with Calponin (Original magnification X200 Calponin).

Tomography (CT) scan showed moderately enhancing left para-orbital soft tissue measuring 12 X 8 mm. There was no evidence of intra orbital extension or bony involvement. Few weeks later, the patient underwent wide excision of the residual suspicious tissue with some normal margin to ensure complete excision of the lesion. The immediate post-operative course was uneventful. The patient showed complete healing with satisfactory cosmetic result when he was seen six months post-surgery (Figure 1b). He was further followed up for a total period of 1 year with no evidence of recurrence. In gross examination, the excised tumor measured 1.8 X 1.5 X 1.0 cm fragment of tan brown soft tissue. Histopathologically, the sections showed interlacing bundles of spindle-shaped cells compatible with immature fibroblasts. The spindle cells had vesicular nuclei and scanty eosinophilic cytoplasm (Figure 2a and b). Frequent mitosis with a low proliferative index and small slit like spaces with scattered red blood cells were also noted (Figure 2c). There were no atypical mitotic figures or cellular atypia. Immunohistochemistry (IHC) staining showed that these cells were positive with smooth muscle actin (SMA), vimentin, calponin, CD68 and CD31 (Figure 2d,e and f). The proliferating cells were negative for desmin (to rule out desmoid tumor), ALK1 (to rule out inflammatory myofibroblastic tumor) and p53. The clinical, histological, and immunohistochemical findings were consistent with NF.

Literature Review

Nodular fasciitis (NF) was first described in 1955 by Konwaler and his coauthors as pseudosarcomatous fibromatosis (Konwaler et al., 1955). They also stressed on the benign nature of NF rather than being a malignant neoplasm (Konwaler et al., 1955). It is commonly seen in the trunk and upper extremities, and rarely involve the orbital region. Font and Zimmerman in 1966 reported the first and largest case series of NF in the ocular adnexal region (Font and Zimmerman, 1966). They presented 10 cases ranging from 3 to 81 years of age. Lesions were reported to occur in the eyelids, periorbital tissue, sub-brow area, in the sub-conjunctiva, and at the limbus (Font and Zimmerman, 1966). Shields reviewed the series with the addition of 13 cases in 2001 (Shields et al., 2001). Since that review, we are aware of 15 more case reports of NF in ocular region in the English-written literature, making our case the 39th reported. Demographic and clinical features of all reported cases of orbital adnexal nodular fasciitis are listed in (Table 1) (Font and Zimmerman, 1966; Shields et al., 2001; Tolls et al., 1966; Levitt et al., 1969; Meacham, 1974; Ferry and Sherman, 1974; Perry et al., 1975; Holds et al., 1990; Vestal et al., 1990; Sakamoto et al., 1991; Kaw and Cuesta, 1993; Meffert et al., 1996; Recchia et al., 1997; Hymas et al., 1999; Ruoppi et al., 2004; Stone and Chodosh, 2005; de Paula et al., 2006; Numajiri et al., 2009; Riffle et al., 2011; Husain et al., 2011; Mukherjee and Mittal, 2012; Gupta et al., 2014; Compton et al., 2014; Phillips and Eiseman, 2014; McClintic et al., 2015; Skippen et al., 2016; Anzeljc et al., 2016; Richards et al., 2016; Massop et al., 2016).

The most common demographic characteristic of NF is the wide age range of occurrence with predominance of adult patients involvement ranging between 20 and 40 years and no apparent gender preference. In this review, the youngest reported case was an 8 months old boy, who presented with an orbital mass simulating a dermoid cyst (Shields et al., 2001). The highest reported age was 81 years with a mean age of about 28 years. Half of the reported cases including ours were in the age group of 30 years or younger (median=30 years). Slight female predominance has been observed in our review with a female to male ratio of 7:6. The right side was more commonly involved in 21/37 of the reported cases where the side of involvement has been recorded. Regarding the exact site of involvement in our review, periocular/orbital involvement such as in our case was more commonly found in 67% (26/39) with deep posterior orbital involvement in 2 cases only out of these. Epibulbar lesions were noticed as a rare entity of orbital NF with only 8 cases reported so far. (Font and Zimmerman, 1966; Tolls et al., 1966; Ferry and Sherman, 1974; Holds et al., 1990; Stone and Chodosh, 2005; McClintic et al., 2015; Massop et al., 2016). Only one case has been reported of epibulbar lesion originating from the limbus in a 3-year-old girl, where the lesion extended to involve the cornea, the iris and the ciliary body (Font and Zimmerman, 1966). Recent article reported the second case of epibulbar NF with corneal involvement (McClintic et al., 2015). Epibulbar NF originating from Tenon's capsule, episclera and sub-conjunctiva has also been reported in literature (Tolls et al., 1966; Ferry and Sherman, 1974; Holds et al., 1990; Massop et al., 2016). Others have presented with a history of floppy eyelid syndrome from frequent and vigorous eye rubbing (Stone and Chodosh, 2005; McClintic et al., 2015).

To the best of our knowledge, there are only 3 reported cases presenting with proptosis (Perry et al., 1975; Compton et al., 2014; Richards et al., 2016). While there has been only 1 reported case originating from the posterior orbit causing optic nerve compression but without proptosis (Gupta et al., 2014). Several cases have been reported to occur during pregnancy, and were thought to be influenced by hormonal changes (Phillips and Eiseman, 2014; Skippen et al., 2016). As in our case, the majority of the reported cases occurred in the periorbital region (Ruoppi et al., 2004; Riffle et al., 2011; Husain et al., 2011; Mukherjee and Mittal, 2012; Phillips and Eiseman, 2014; Richards et al., 2016).

In the literature, prior history of trauma was found in 10% to 15% of the cases (Font and Zimmerman, 1966). NF should be included in the differential diagnosis of rapidly growing, soft tissue mass in the periorbital and orbital regions to avoid misdiagnosis of a malignancy and

Table 1. Reported Cases of Nodular Fasciitis in Ocular Adnexal Region

Case	Authors (Year)	Age (y)	Gender	Side	Location
1	Font & Zimmerman (1966)	3	F	R	Limbus
2	Font & Zimmerman (1966)	52	F	L	Periorbital
3	Font & Zimmerman (1966)	25	F	L	Epibulbar
4	Font & Zimmerman (1966)	46	M	R	Eyelid
5	Font & Zimmerman (1966)	28	M	NR	Anterior orbit
6	Font & Zimmerman (1966)	30	M	L	Eyelid
7	Font & Zimmerman (1966)	81	F	R	Medial canthus
8	Font & Zimmerman (1966)	61	F	R	Medial canthus
9	Font & Zimmerman (1966)	21	F	L	Lateral canthus
10	Font & Zimmerman (1966)	37	F	L	Eyebrow
11	Tolls et al (1966)	43	M	L	Epibulbar
12	Levitt et al (1969)	14	M	R	Anterior orbit
13	Meacham (1974)	59	M	L	Eyebrow
14	Ferry & Sherman (1974)	47	F	L	Epibulbar
15	Perry et al (1975)	34	F	L	Anterior orbit
16	Holds et al (1990)	3	F	R	Epibulbar
17	Vestal et al (1990)	11	M	L	Eyelid
18	Sakamoto et al (1991)	7	F	R	Medial canthus
19	Kaw & Cuesta (1993)	10	F	L	Anterior orbit
20	Meffert et al (1996)	26	M	R	Eyelid
21	Recchia et al (1997)	3	F	L	Anterior orbit
22	Hymas et al (1999)	14	M	NR	Eyelid
23	Shields et al (2001)	<1	M	R	Anterior orbit
24	Ruoppi et al (2004)	1	F	L	Periorbital
25	Stone & Chodosh (2005)	53	M	R	Epibulbar
26	De Paula et al (2006)	3	M	R	Eyelid
27	Numajiri (2009)	53	M	R	Eyelid
28	Riffle et al (2011)	45	M	R	Periorbital
29	Husain et al (2011)	30	F	R	Periorbital
30	Mukherjee & Mittal (2012)	9	F	L	Periorbital
31	Gupta et al (2014)	16	F	R	Posterior orbit
32	Compton et al (2014)	1	M	R	Posterior orbit
33	Phillips & Eiseman (2014)	25	F	R	Periorbital
34	McClintic et al (2015)	38	M	R	Epibulbar
35	Skippen et al (2016)	31	F	R	Medial canthus
36	Anzeljc et al (2016)	38	F	R	Eyelid
37	Richards et al (2016)	12	M	R	Periorbital
38	Massop et al (2016)	32	F	L	Epibulbar
39	Present case (2016)	35	M	L	Periorbital

NR: not recorded

unnecessary aggressive treatment (Tolls et al., 1966; Kim et al., 2005). The clinical differential diagnosis of NF includes fibrous histiocytoma, malignant fibrous histiocytoma, sarcoma, fibromatosis, fibroma, neurofibroma, neurilemmoma, and leiomyoblastoma (Vestal et al., 1990).

Imaging studies show no distinctive features to differentiate nodular fasciitis from other solid masses. On computed tomography or magnetic resonance imaging, NF typically show lesions enhancement with contrast and often show local bony erosion, with no pathognomonic imaging feature (Kim et al., 2005).

The histopatholgic features of NF can raise the concern of rhabdomyosarcoma or other soft tissue sarcoma due to their abundant spindle cells and high mitotic activity. Lesions are often showing plump stellate or spindle shaped fibroblasts usually arranged in parallel bundles and extending in all directions, which is similar to their appearance in tissue culture. Fibroblasts are intermixed in myxoid ground substance (Shields et al., 2001; Mukherjee and Mittal, 2012). Immunohistochemistry can aid in differentiating NF from other soft tissue lesions. Cells in NF express muscle specific actin, smooth muscle actin, and vimentin (Shields et al., 2001; Mukherjee and Mittal, 2012). On the other hand, the spindle cells in NF do not express desmin, keratin, ALK1 and S-100 (Kayaselçuk et al., 2002).

Surgical excision is the modality of choice for NF involving ocular adnexa. Local recurrence is very rare following complete excision (Shields et al., 2001; Vestal et al., 1990). Recently, intra-lesional steroid injection use has been also reported (Husain et al., 2011).

CONCLUSIONS

This is the first case of nodular fasciitis involving the periorbital region of an adult in the Middle East to the best of authors' knowledge. In addition, we have reviewed all the reported cases of NF in ocular adnexal region in the English-language literature. We can conclude that NF is a rare, benign fibroblastic proliferation that should be included in the differential diagnosis of rapidly growing, soft tissue mass in the periorbital and orbital regions. NF occurs within a wide range of age (8months to 81years) with a median age of 30 years). Slight female predominance is observed with a F:M ratio of 7:6. The right side is more commonly involved (in 21/37). Periocular/orbital involvement is more commonly found (67%). Although rare, these lesions are clinically and pathologically concerning for malignancy.

ACKNOWLEDGEMENT

This work was supported by the College of Medicine Research Center, Deanship of Scientific Research, King Saud University, Riyadh, Saudi Arabia

Declaration Statement

Authors have no conflict of interest of any type related to this work. No funding was received from individuals or corporations in relation to this paper.

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