Periorbital nodular fasciitis arising during pregnancy

Brandon N Phillips, Andrew S Eiseman

Nodular fasciitis (NF) is a benign proliferation of fibroblasts and myofibroblasts that rarely occurs in the periorbital region. We report what we believe to be the first case of periorbital NF associated with pregnancy. A case of intravascular fasciitis, a NF variant, has been reported during pregnancy, but it was not located in the periorbital region. A weak presence of estrogen receptors has been reported in NF. This may make it more susceptible to the hormone-related changes during pregnancy and contribute to the development of the lesion by stimulating fibroblasts and smooth muscle cell types. Although rare, NF should be considered in the differential diagnosis of periorbital soft-tissue masses arising during pregnancy.

Key words: Nodular fasciitis, periorbital, pregnancy, sarcoma

Nodular fasciitis (NF) is a benign proliferation of fibroblasts and myofibroblasts that is characterized by a solitary, firm, rapidly growing nodule. It occurs predominately in the upper extremities and trunk, but it has also been identified in the head and neck region in 13–20% of reported cases.[1] Periorbital involvement is even less common although involvement of the eyebrow, eyelids, and ocular surface have been described.[2-4] Lesions are typically self-limited but can be mistaken for a sarcoma due to its rapid growth. NF can occur at any age but young adults are most commonly involved, affecting males and females equally. NF does not appear to be associated with any systemic manifestations, although a case of intravascular fasciitis, a variant of NF, was reported in a pregnant woman.[5] A literature review could find no reports of any other cases of NF in pregnancy. We report what we believe to be the first case of periorbital NF associated with pregnancy.

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Ophthalmology Service, Walter Reed Army Medical Center, Washington DC, USA

Correspondence to: Dr. Brandon N. Phillips, Ophthalmology Service, Walter Reed National Military Medical Center, 8901 Wisconsin Avenue, Bethesda, MD 20889, USA. E-mail: brandon.n.phillips@us.army.mil

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Case Report

A healthy 25-year-old Caucasian female presented 2 months postpartum complaining of a mass above her right eye. She first noticed it when she was 6 months pregnant and it had progressively enlarged. The patient reported mild tenderness to palpation. The forehead and scalp had normal sensation to touch. It was the patient's first pregnancy and she was breastfeeding. The patient denied a history of trauma. Examination revealed a freely mobile 1 × 1 cm firm, solitary nodule above the superomedial aspect of the right orbital rim. The surrounding tissue was not adherent and the overlying skin was unremarkable. There were no other lesions or lymphadenopathy. A computed tomography (CT) scan examination of the orbits showed a rounded, soft-tissue mass in the subcutaneous soft tissue at the right supraorbital notch [Fig. 1]. There was no associated osseous defect and no intraorbital, sinus or other extension identified.

The nodule was completely excised and the surgical site closed primarily. Histopathological examination revealed a slightly irregular unencapsulated mass with monomorphic spindle-shaped cells. They were arranged in interlacing fascicles with occasional storiform pattern in a myxoid stroma [Fig. 2]. Mitoses were infrequent and scattered throughout the lesion with no atypical mitoses. There were numerous fine capillaries with scattered collections of extravasated lymphocytes and red blood cells present. The tumor cells stained positive for smooth muscle actin (SMA) and vimentin. They stained negative for S100 and estrogen and progesterone receptors.

Discussion

Periorbital NF is a relatively rare lesion. The etiology remains unknown although trauma and inflammatory reactions have been blamed even though limited evidence substantiates this. Periorbital NF can be very concerning since its rapid onset and growth may simulate a malignant process. Although periorbital lesions have similar histological features when compared to NF identified at other anatomic sites, the lesions tend to be smaller presumably due to the lack of abundant subcutaneous fat and earlier patient presentation.[1] Local marginal excision is the preferred treatment although partial resection may be adequate. Recurrence of the excised lesion has been documented, but it is so uncommon that it has been suggested that recurrence of a lesion initially diagnosed as NF should lead to a careful reevaluation of the original diagnosis.[6]

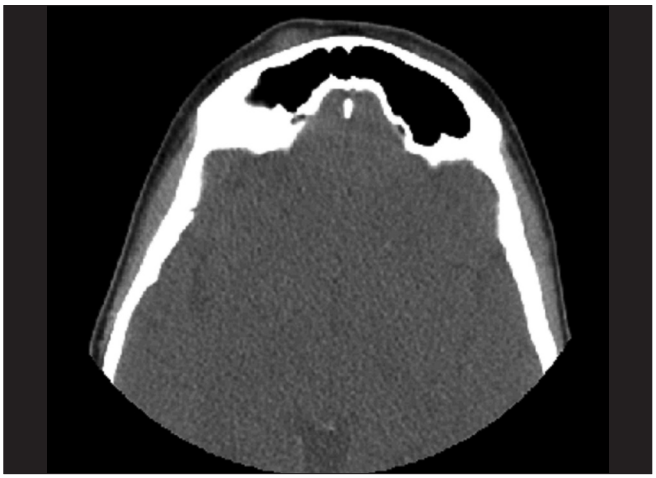

Figure 1: A rounded subcutaneous soft-tissue mass at the right supraorbital notch. There is no associated osseous defect or extension identified

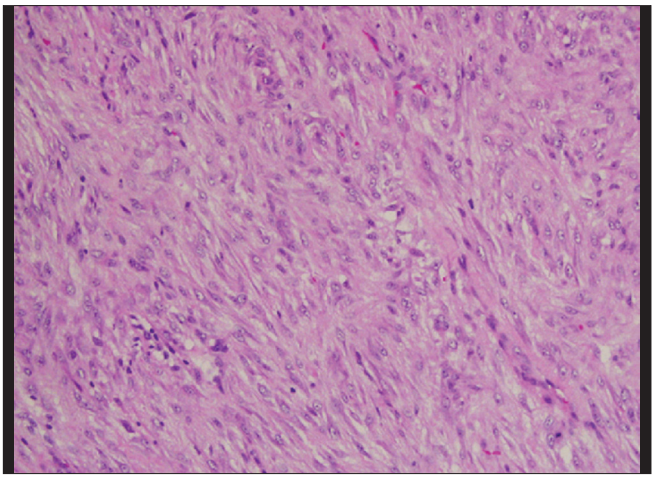

Figure 2: Bland plump spindled cells in a loose myxoid background with extravasated red blood cells and scattered chronic inflammation, (H and E, ×200)

Ophthalmologists are unlikely to consider NF in their differential of lesions in the periorbital region. Clinically, the differential diagnosis may include fibroma, solitary fibrous tumor, dermatofibroma, fibrosarcoma, neuroma, neurofibroma, lipoma, granuloma, dermoid, and epidermal inclusion cyst. The clinical and imaging features of NF are not pathognomonic and this can lead to potential diagnostic confusion. NF is a benign lesion that does not metastasize. Histopathologic examination and immunohistochemistry are usually required for the precise diagnosis. Despite the absence of cellular atypia, NF can be confused with a spindle cell sarcoma because of its infiltrative quality, rich cellularity, and variable mitotic figures.[7] These include fibrosarcoma, neurofibrosarcoma, myofibroblastic sarcoma, and osteosarcoma.

As a spindle cell tumor, NF stains positive for vimentin and variably for SMA. Similar to our case, NF in the head and neck region more commonly stains diffusely for SMA than lesions of the trunk and extremities, suggesting stronger expression by myofibroblasts. NF does not stain positive for desmin, keratin, or S-100 protein.[6] A weak presence of estrogen receptors was reported in a series with three out of four cases of back and upper extremity NF, demonstrating focal cytoplasmic positivity for estrogen receptors.[6] A case of intravascular fasciitis, a NF variant, was discovered during pregnancy and the authors hypothesized that the hormone-related changes during pregnancy may have contributed to the development of the lesion.[5] They reasoned that estrogen is known to stimulate fibroblasts and smooth muscle cell types and has been implicated in other fibroproliferative disorders such as carpal tunnel syndrome. They did not report staining their specimen for estrogen receptor antibodies.

Since our patient developed NF during pregnancy without a history of trauma, we also reasoned that hormone-related changes may have contributed to the development of the lesion by inducing smooth muscle and fibroblast proliferation. The lesion stained negative for estrogen and progesterone receptors, but this can sometimes be due to poor fixation and embedding procedures.[8] Although rare, NF should be considered in the differential diagnosis of periorbital soft-tissue masses arising during pregnancy.

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