

## MULTIPLE NODULAR FASCIITIS PRESENTING INITIALLY AS A CHEST WALL TUMOR

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Nodular fasciitis is a benign proliferation of myofibroblast often misdiagnosed as a sarcoma of soft tissues. It is characterized by rich cellularity, mitotic activity and rapid growth. It is usually solitary, and occurs in the head and neck in childhood, and upper extremities in adulthood. We report the case of a four-year-old child who developed multiple lesions in the head, neck and trunk. Nodular fasciitis was diagnosed in this reported case by incisional biopsy.

### Case Report

A four-year-old Sudanese boy presented with a two-week history of a painless scapular mass, which was progressively increasing in size and was associated with moderate restriction of right arm movement. His father denied any history of trauma. The patient had a history of bilateral halux valgus, which had been corrected two years previously. He was a product of a spontaneous vaginal delivery and had no family history of a similar problem. The result of clinical examination showed a 5x10 cm firm mass at the angle of the right scapula fixed to underlying muscle. The area was not tender to palpation, had no warmth or erythema, and the overlying skin was freely mobile. There was limitation of abduction up to 70°, and restriction in bending forward due to a bulky right paraspinal muscle. Initial laboratory findings revealed the following: WBC count, 8.2x10/L; ESR, 7 mm in the first hour; creatine kinase, 169 U/L (normal, 25-200 U/L); alkaline phosphatase, 455 U/L (98-279 U/L); and calcium level of 10.46 mg/dL (8.1-10.4 mg/dL).

The radiological evaluation of the patient included a chest x-ray (Figure 1), which showed right-sided chest wall swelling mainly at the scapular region. The opacity was homogenous, with no calcification or bony lesion.

Enhanced CT scan showed slightly infiltrating mass of soft tissue density among muscles of the right shoulder and chest wall. Fine-needle aspiration was inconclusive. The patient was taken to the operating room, and incisional biopsy was performed under general anesthesia. At surgery, there was no line of demarcation between the subcutaneous tissue, tumor and underlying muscle. Therefore, a wedge biopsy was taken, including subcutaneous tissue and muscle. Closure was performed in the usual fashion.

The patient was discharged home while awaiting histopathology report. Microscopic examination of the biopsy demonstrated a sparsely cellular lesion, composed of immature, plump and slender fibroblasts in a loose background, and containing extravasated red blood cells and rare inflammatory cells, particularly mast cells. The fibroblasts were haphazardly arranged, relatively uniform in size and shape, and had oval pale staining nuclei, with prominent nucleoli and rare mitotic figures. These cells resembled fibroblasts seen in tissue culture (Figure 2).

Residual skeletal muscle fibers were seen within the lesion, which indicated infiltration of the surrounding skeletal muscle. These microscopic features were consistent with nodular fasciitis.

The child was seen in the outpatient clinic one week after the operation. Clinical examination showed regression of the scapular mass, but the patient subsequently developed multiple masses over a period of three months, initially at the angle of the left scapula, then nape of the neck and occiput of the skull. Follow-up radiological examination of the chest showed a new bone formation at the right and left chest wall and left cervical region (Figure 3).

The patient was followed up for the next 18 months, and during this period he was treated conservatively with limb physiotherapy. Although he was able to cope with daily activities, the movement of both arms, neck and back was progressively restricted.

### Discussion

Nodular fasciitis (NF) is a benign proliferation of myofibroblast,<sup>1,2</sup> which may be misdiagnosed as a sarcoma due to its rich cellularity, mitotic activity and variant

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morphologic pattern.<sup>1,3</sup> It commonly occurs in the head and neck area in the pediatric population, while upper extremities are the favored location in adults.<sup>4,5</sup> Other rare locations reported have been in the buccinator muscle,<sup>1</sup> breast,<sup>6</sup> parotid gland,<sup>2</sup> auricle,<sup>7</sup> and the dermis.<sup>8</sup>

NF commonly occurs in young adults. Samaratunga et al. reported 33 cases of NF, and 44% of these were between the ages of 20 and 29 years, with only one case below the age of nine years.<sup>3</sup> NF is of unknown etiology, but local injury or an inflammatory process are believed to be the triggering factors.<sup>1,3,4</sup> The important clinical features of NF are its history of rapid growth and small size. In our reported case, the interesting points were the occurrence of the disease in a four-year-old child and its multiplicity, as NF is generally solitary.<sup>2</sup> There have been few previous reports describing multiple lesions of NF.<sup>9,10</sup> Other lesions that subsequently developed at the patient's left scapula and neck, similar to the initial lesion at the right scapula, were lesions of NF. A biopsy was not taken, as the patient's father was reluctant for a second biopsy to be taken from the newly developed lesions.

The radiological examination in Figure 3 demonstrates ossification in all lesions, denoting the same pathology. Kaw and Cuesta reported the diagnosis of NF by fine-needle aspiration (FNA) cytology preoperatively.<sup>5</sup> Although FNA was inconclusive in our reported case, the incisional biopsy confirmed the diagnosis with a typical microscopic description of NF. The patient was subjected to incisional biopsy because our initial clinical diagnosis was rhabdomyosarcoma, and a biopsy was required preoperatively to justify aggressive resection.

There are many other lesions which should be considered in the differential diagnosis of a mass at the back and around the scapula. These include rhabdomyosarcoma, fibrosarcoma, neurofibrosarcoma, osteosarcoma, Askin's tumor, and fibroma. In their study, Zuber and Finley provided a table of the list of lesions to be considered in the differential diagnosis.<sup>4</sup> Most of these tumors can be excluded microscopically. Red blood cell extravasation, keloid type collagen, and lack of pleomorphic cells are the main features favoring a diagnosis of NF.<sup>11</sup> However, one might anticipate a close resemblance of fibrosarcoma and neurofibrosarcoma to NF. In contrast to NF, fibrosarcoma is a deep-seated and highly cellular neoplasm. The cells are arranged in characteristic "herringbone" pattern, with hyperchromatism of nuclei and greater degree of mitotic activity. Similarly, proximity to a major nerve trunk, high cellularity and typical serpentine cell arrangement in palisades or whorls, or large gaping vascular spaces, are useful distinguishing criteria of neurofibrosarcoma.<sup>11,12</sup>

Another interesting point to mention in NF is the presence of infiltrative margin,<sup>11</sup> and the possibility of recognizing residual muscle fibers,<sup>12</sup> as found in our reported case. It does not imply malignant nature but

rather the ability of NF to simulate infiltrative malignant process.

Most of the reports we reviewed diagnosed the lesion as NF after local excision, and therefore, we advise surgical resection as the treatment of choice. We did not proceed to this option in our reported case because of the multiplicity of the lesion and the involvement of different muscle groups, including vital muscles of the neck and back.

Samaratunga and colleagues reported three cases of ossifying fasciitis.<sup>3</sup> They were morphologically typical lesions of NF, and were associated with metaplastic bone formation, however, our reported case demonstrated that ossification of the lesion is the subsequent progression of the disease. Daroca et al. explained the link between NF and myositis ossificans as due to the presence of focal metaplastic bone formation in NF.<sup>13</sup> All the reported cases by Samaratunga were well and alive three months to 13 years after simple excision, and none had recurrence or metastasis.<sup>3</sup> Our reported case has been followed up for 18 months after the diagnosis and subsequent supportive therapy, however, he has been developing progressive restriction of movement of upper arms, neck and back. Spontaneous regression of incompletely excised lesion of NF has been noted,<sup>9</sup> and almost all reports advocate no further treatment once the lesion is surgically excised.

In summary, nodular fasciitis is one of the differential diagnoses of a mass arising from the chest wall. Physicians and surgeons who are facing a chest wall tumor should be aware of this differential diagnosis and should work closely with the pathologist in confirming the diagnosis. Further follow-up of our reported case will be continued for a longer period to determine the prognosis of the unresected lesion of NF.

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