

A Case Of Fibromatosis In A 45 Days Old Infant

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Citation

R Gupta, I Wani, N Gupta, S Gupta, H Singh, D Singh. *A Case Of Fibromatosis In A 45 Days Old Infant*. The Internet Journal of Orthopedic Surgery. 2008 Volume 11 Number 1.

Abstract

Fibromatosis lesions are rare neoplasms arising from fascial or deep musculo-aponeurotic structures that display local aggressiveness but no propensity to metastasise. These fibrous tumours occurring in infants and children range from sparsely cellular proliferations of spindle shaped cells (designated as fibromatosis) to richly cellular lesions indistinguishable from fibrosarcomas occurring in adults. These fibromatosis lesions presenting in infants and children although usually present solitary, they may be multifocal involving any organ. Solitary lesions are benign. Fibromatosis is synonymous to extra-abdominal desmoid tumor 3. The most common sites of extra-abdominal desmoid tumor are shoulders followed by chest, back, thigh, head and neck 2.

Overall, desmoid tumors are reported to account for 0.03% of all neoplasms 4. The incidence of the fibromatosis occurring in the extremities and trunk is almost half of all the desmoid tumors. Fibromatosis has a female preponderance.

CASE REPORT

A 45 days old female infant was brought to the hospital with swelling in the right thigh (Fig.1).

Figure 1

Figure 1: showing photograph of a patient with illdefined swelling in the distal part of right thigh.



Parents of the baby had noticed the swelling when she was 11 days old. There was no history of fever, pain or birth

trauma. The swelling was diffuse extending from lower end of femur up to the midshaft whose margins were indistinct on inspection but were well defined on palpation. Temperature over the swelling was normal. There was no tenderness. Shape was fusiform and was firm to hard in consistency. It was non mobile, overlying skin and surrounding muscles were free.

Lymphnodes in drainage area were not palpable. Distal neurovascular status appeared to be normal. Radiograph when patient was 11 days old did not show any abnormality (Fig.2)

Figure 2

Figure 2: showing no radiographic abnormality when patient was 11 days old



However at 45th day of life, there was a circumferential overgrowth of radio opaque tissue which covered the normal bone like a shell.

Figure 3

Figure 3: showing circumferential overgrowth of radioopaque tissue



Cortices of underlying bone were intact. FNAC and Tissue Biopsy showed features suggestive of fibromatosis.

DISCUSSION

This case of fibromatosis surrounding femur was reported for the first time in our hospital. As we know that incidence of this case is very low, we decided to report this case. Biologically, deep seated fibromatoses lie in the borderland between nonaggressive fibrous tumors and low-grade fibrosarcomas. On the one hand, they commonly present as large infiltrative masses that frequently recur after incomplete excision; on the other hand, they are composed of banal of well differentiated fibroblasts that do not metastasize but has high incidence of recurrence. We have performed marginal excision of the growth and the case is under follow up. Leithener et al¹, in 2004 reported 72% recurrence in marginal or intralesional surgical procedures as compare to 15% recurrence in wide resection.

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