ABSTRACT

Fibrous Hamartoma of Infancy is a rare juvenile soft tissue tumour that may be confused with a sarcoma, as occurrences are known to occur. These benign growths may recur and caution is advised since the microscopic pattern may be misinterpreted as a low grade sarcoma. Complete excision with adequate margins is required, as spontaneous regression is not to be expected.

Keywords: Fibrous Hamartoma, Tumour, Cells, Stroma.

FIBROUS HAMARTOMA OF INFANCY

Fibrous hamartoma of infancy [1] is a rare, and unusual soft tissue growth occurring exclusively in infants less than 30 months. The microscopic picture may be confused with a low-grade fibrosarcoma [2] however, the histologic pattern need not cause confusion once the pathologist is familiar with this distinctive neoplasm. To our knowledge the occurrence of this rare tumour on the lower extremity has been reported; only a few times and only one case in the original series had two recurrences.

Although Enzinger [3] named the tumour, Reye [4] first brought attention in 1956 to unusual subdermal tumors of male infants with a common histologic pattern. He believed they represented a reparative response, and that they were wholly benign.

In 1965 Enzinger collected thirty cases submitted to the A.F.I.P. for consultation. Submitting diagnoses ranged from fibroma to embryonal rhabdomyosarcoma. All patients first presented under the age of thirty months; all tumors were located above the groin; three recurred once. A single case had two recurrences (the site is not specified). Enzinger noted that cases 28 and 29 of Stout’s series on Juvenile Fibromatoses [5,6] satisfied the criteria of fibrous hamartoma of infancy.

A twenty-eight-month old boy was admitted to the Berkshire Medical Center because of a swelling on the plantar left foot of one month’s duration. The boy had no other apparent physical problems. Family history was not contributory, and trauma was not involved. Examination revealed a soft tissue mass situated along the medial aspect of the left foot 2 cm distal to the medial malleolus, measuring 3.5 x 3.5 x 2.0 cm. The mass seemed to extend across the transverse plantar arch to reach within 1.5 cm of the lateral plantar margin. It had an indurated, rubbery consistency. No pain was elicited by compression and skin changes were present resembling parchment. Roentgenograms of the feet showed
minimal decalcification of the cuneiforms but no evidence of erosion or new bone growth.

A biopsy was performed (Figure 1). Microscopy revealed a tumour composed of bundles of dense interlacing fibrous tissue containing fat and islands of loose, whorled, mesenchymal-like cells in a mucoid-appearing matrix. The fibrous component was characterized by a rippled, swirling pattern without evidence of mitosis. Fatty elements resembled mature adipose tissue. Islands of loose mesenchymal-like cells stood out as translucent bluish foci contrasted to the dense eosin-staining fibro collagenous stroma. Masson's trichrome Mucicarmine staining induced a pale stain yielded a rich green fibrous stroma. Pink colour in the matrix of the mesenchymal component. Reticulum stains demonstrated fine wavy fibrils in these foci. A diagnosis of fibrous hamartoma of infancy was established, and the entire tumour was therefore resected. The surgical specimen measured 7 x 2 x 1 cm and was lipomatous-appearing with fibrous areas, and partially encapsulated. (Figure 2) A pedicle of tumour was found to extend between the plantar fascia and the inferior surface of the os calcis to reach the lateral border of the foot. The dissection was performed en toto. Neurovascular structures could not be identified. Measurement, including the pedicle, was 7 x 2 x 1 cm. The surface was gray, firm, and glistening.

Figure 1. Microscopic picture characterized by bundles of interlacing fibrous tissue containing fat and islands of mesenchymal-like cells. (430x)

Figure 2. Gross appearance showing islands of "myxoid" tissue interspersed by fibrous tissue.
Eight months later the tumour recurred, heralded by discomfort in the arch of the foot. The mass was again excised. The entire specimen measures 4 x 3 x 2 cm. It was noted by the surgeon to involve the posterior tibial neurovascular bundle. The histologic pattern was identical to tile previous excision and neural elements did not predominate. Two years following the initial excision, and sixteen months after the second excision, there was a second recurrence. However, this time there was no pain, nor was walking affected. Noted at surgery was the encapsulation of the tumour and the extension across the foot to the os calcis; this portion of the tumour was markedly adherent to the lateral aspect of the tuberosity of the os calcis, and was removed together with periosteum. The resected tumour measured 6 x 3 x 3 cm. Also involved were the flexor hallucis longus and the medial plantar nerve. Fibrous hamartoma of infancy may not be suspected when a soft tissue neoplasm involves the foot. However, biopsy is not difficult to interpret once the pathologist is familiar with the distinctive histology. The natural history of the tumour seemingly is benign and local resection has been the treatment of choice. An unusual soft tissue tumour, fibrous hamartoma of infancy, is presented. As described first by Reye, and subclassified by Enzinger, the tumour is thought to be benign, although recurrences are reported. This case demonstrated the unusual location, and the nature of recurrence. The pathologic diagnosis is not difficult once the distinctive pattern is recognized.

REFERENCES
2. Ibid. p. 242
3. Ibid. p. 248