Acta orthop. scand. 47, 370–372, 1976

INTRAOSSEOUS NEUROFIBROMA OF THE CLAVICLE

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A case of an intraosseous neurofibroma of the medial end of the clavicle, treated surgically, is reported. The relevant literature is reviewed.

Key words: neurofibroma; clavicle

Accepted 21.iii.76

Primary nerve tumours are extremely rare. Fawcett & Dahlin (1967) found seven cases in their series of 3987 primary bone tumours at the Mayo Clinic and were able to collect another 31 cases after a review of the literature. No case of intraosseous neurofibroma of the clavicle either alone or in association with multiple neurofibromatosis has been reported in the literature reviewed. This prompted us to report this rare case of intraosseous neurofibroma.

CASE REPORT

V.L., a 34-year-old Hindu male, was admitted with complaints of progressive pain and swelling over the medial aspect of the right clavicle which had lasted for the previous 4 to 5 months. There was no history of injury, fever or pain or associated swelling in other parts of the body.

On examination, a diffuse, firm oblong swelling 7.5 cm × 5 cm in size extending from the junction of the medial third of the clavicle to the suprasternal notch was seen. No pressure effects were detected either proximally or distally.

There was no evidence of stigmata or appearances suggestive of multiple neurofibromatosis. A skiagram of the clavicle showed an osteolytic lesion of the medial third of the clavicle, with no incipient new bone formation (Figure 1). A scalloped appearance of the normal medial end of the clavicle was seen. Skiagrams of the chest and cervical spine and a blood examination were within normal limits.

The mass was explored by an incision 10 cm long from the medial third of the clavicle to the suprasternal notch. Extra-periosteal resection of the clavicle 4 cm lateral to the palpable mass was done. Haemostasis was maintained and the wound closed after approximating the pectoralis major and the sternomastoid. The patient made an uneventful recovery. At follow-up after 1 year there was no recurrence of the tumour.

Pathology

Gross: The firm to hard irregular growth measuring 1.5 × 3 × 6 cm in size was attached at one end to a 4 cm long piece of bone. The outer layer was white and streaked.

Microscopic: Most of the sections showed a uniform picture of well-formed mature connective tissue with thin elongated nuclei with pointed ends. In these regions there was no pleomorphism etc. However, in two of the sections towards one edge there were sufficiently large areas showing a variable degree of cellular pleomorphism and these areas also showed a ponceau-positive cytoplasm. There was no evidence of bony or cartilagenous tissue. The overall histological appearances were in favour of a neurogenic tumour of very low malignancy or a fibroblastic tumour.

DISCUSSION

Miller & Kashara (1963) and Sherman (1963) demonstrated both myelinated and non-myelinated nerves entering vari-

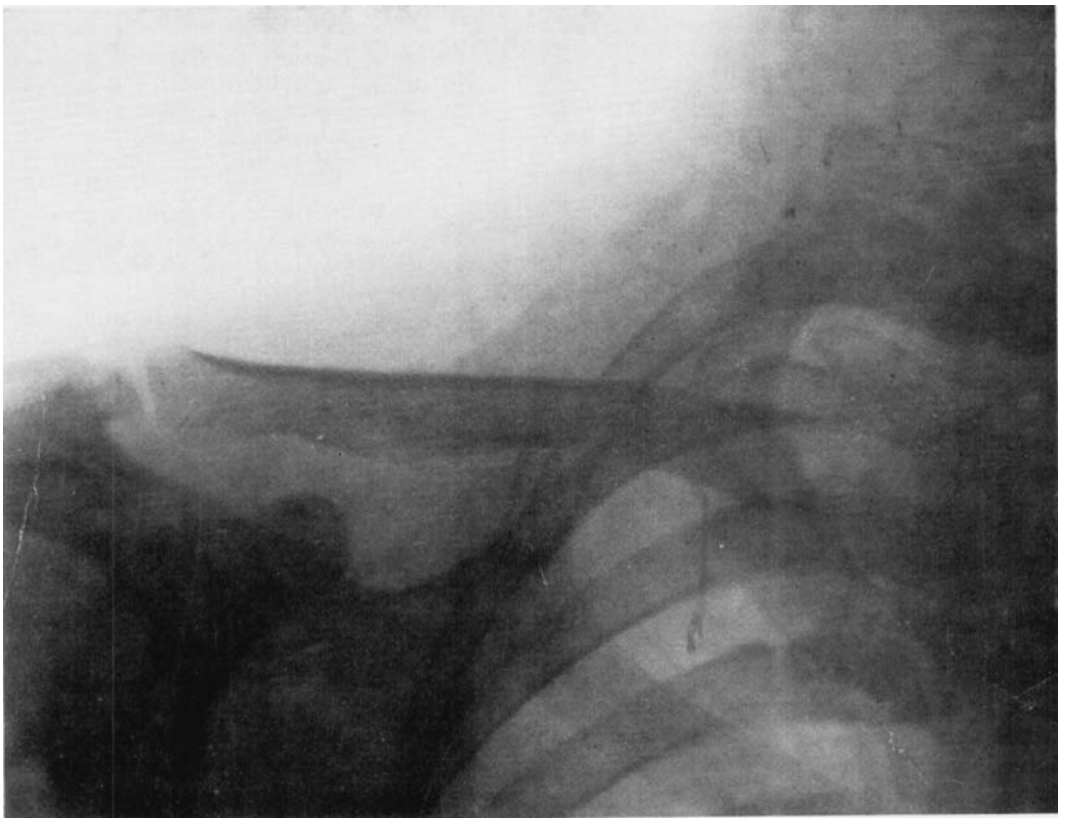

Figure 1. Osteolytic lesion of the medial third of the right clavicle. There is no evidence of bone formation.

ous foramina in bones often in association with blood vessels. These nerves may serve as the tissue from which neurofibromas arise. Most of the cases of intraosseous nerve tumour reported have been related to the proximity of nerves and nutrient foramina. Fawcett & Dahlin (1967) reported seven cases of intraosseous neurogenic tumour, four in the mandible and one in each of the third rib, the scapula and the femur. He reviewed a total of 38 cases but there were none involving the clavicle. Most intraosseous neurilemmomas have occurred in the mandible; the long intraosseous course of the relatively long mental nerve is probably the reason for the occurrence of neural tumours in that bone. Other nerve tumours at other sites have been demonstrated close to the nutrient foramina. Brooks & Lehman (1924) suggested that with the development of a neurofibroma in the periosteum, there is a certain amount of reaction and bone destruction and regeneration follows. This results in a certain type of subperiosteal or cortical cyst. Cases of this type have been reported in the literature (Henseley 1953, Sane et al. 1971). In the present case, however, there was no evidence of periosteal reaction. Hence it seems most probable that the tumour arose from the nerve entering along with the nutrient vessels.

The differential diagnosis in cyst-like lesions of this type includes other benign fibrous lesions. Neurilemmoma, however, lack the metaplastic osteoid of fibrous

dysplasia, the giant cells of fibroma of bone and the whorled dense fibrous tissue of the desmoplastic fibroma. In the present case, there was initially difficulty in arriving at a diagnosis but a detailed examination of the blocks of the tissue coupled with systematic exclusion of other lesions pointed to the diagnosis of neurofibroma.

ACKNOWLEDGEMENTS

I am grateful to Dr. N. S. Pathania, Principal and Dean, Govt. Medical College and S.M.G.S. Hospital, Jammu, for permission to publish this case and I am indebted to Dr. Dutta of the Post Graduate Institute, Chandigarh, and Dr. S. Sriramachari of the Indian Registry of Pathology, Safdarganj Hospital, New Delhi, for histopathological examination.

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