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CALCIFYING FIBROUS PSEUDOTUMOR : CASE REPORT

KALSİFİYE FİBRÖZ PSÖDOTÜMÖR : OLGU SUNUMU

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SUMMARY

This report concerns the finding of a clinically malignant mass in the subcutaneous tissue of the left supraclavicular region of a 6-years-old boy. Microscopic examination proved jt to be an unusual lesion recentfy entithd "catctfying fibrous pseudotumor (CFP)". İt is a rare benign tumour characterised by the presence of abundant hyalinized collagen with psammomatous or dystrophic calcification and scarce lymphoplasmacytic infiltrate. Total excision was performed. This procedure is thought to be necessary to preclude local recurrence.

ÖZET

Bu olgu sunumu 6 yaşında bir erkek çocuğunda sol supraklaviküler bölgede subkutan dokuda saptanan klinik olarak malign kitleyi içermektedir. Mikroskobik inceleme son zamanlarda kalsifiye fibröz psödotümör olarak tanımlanan nadir bir lezyonu ortaya koydu. Çok fazla miktarda hyalinize kollagen, psammamatöz ya da distrofik kalsifikasyon ve seyrek lenfoplazmositik infiltrasyonla karakterize nadir benign bir tümördür. Total eksizyon uygulanmıştır. Bu girişim lokal rekürrensi önlemek için gerekli görülmüştür.

CASE

A five years old boy was seen in April 2000 because of a mass in the left supraclavicular region noticed two months before. He had slight pain with palpation and had noticed recent increase in size of the mass. Physical examination revealed a five by three centimetre, hard, mobile, tender mass in the subcutaneous tissue of the neck. İt was suspected to be a lymphoproliferative disorder and an incisional biopsy was performed in May 2000. The biopsy specimen consisted of a small lymph node and a hyalinized fibrosclerotic tissue fragment with it. A distinctive feature of this tissue was the presence of scattered calcium deposits. Reactive follicular hyperplasia in the excised lymph node and dystrophic calcification on the other biopsy material were reported and also investigation of the metabolic disorders was recommended. The patient was followed up for ten months. Because the mass continued to increase in size, 11 months later it was completely excised.

On gross examination, the lesion was found tc be a six by five by three centimetre, firm, solid, lobular, well-circumscribed but unencapsulated mass. The cut surface was grey-white with a gritty texture. Meanwhile the first

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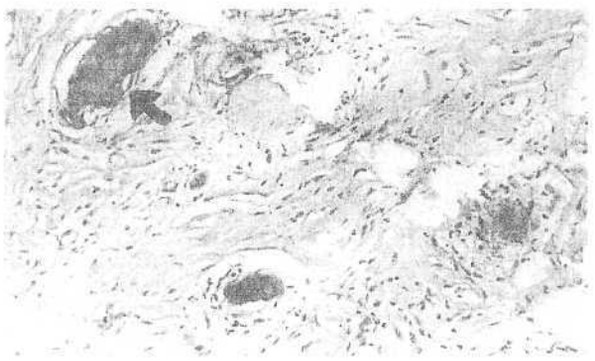
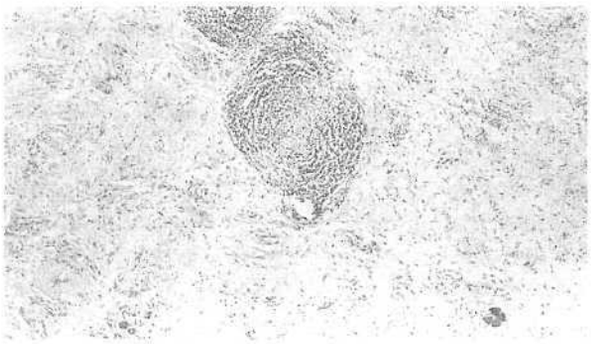
biopsy specimen was reviewed. The second biopsy material and the little soft tissue fragment of the first biopsy material were similar microscopically and were composed of mostly dense hyalinized collagenous tissue interspersed with benign spindle cells, A sparse benign inflammatory infiltrate of plasma cells and lymphocytes was present scattered among the collagen bundles. The diagnostic feature of these lesions was the presence of scattered calcifications. Some of this had a laminated appearance typica! of psammoma bodies. Others were dystrophic in character (Fig 1). Occasional foreign body giant cells and lymphoid aggregates were noted (Fig 2)



Figüre 1. The lesion was characterised microscopically by dense bands of collagen with scattered inflammatory cells and several calcifications. One of thern concentrically laminated resembling psammoma bodies (arrow). Others are dystrophic (HEX40Q).



Figüre 2. Elsewhere, there were small collections of lymphocytes forming germinal centres (HEX 100).

DISCUSSION

CFP is a rare, benign, tumorlike lesion that is characterised by fibrotic proliferation, infiltration of inflammatory cells and calcification. As far as we know, in 1988, Rosenthal and Abdul-Karim (1) presented two similar lesions as "fibrous tumour of childhood with psammoma bodies". Fetsch and colleagues (2) from the Armed Forces Institute of Pathology, in 1993, reported only ten patients who had what was described as a "calcifying fibrous pseudotumour". Recently, this seccnd designation is preferred because the lesions afflict a broader age range than initially appreciated and because it reflects that the underlying process is most likely fibro-inflammatory and reactive. The adjective "calcifying" has been used as a substitute for psammoma bodies because the mineralisation is sometimes of the dystrophic type. The ultrastructural examinations of the CFP revealed the deposition of calcium along the extracellular collagen fibers as dystrophic type as well as intracellular calcification in residual bodies as psammomatous type (1-3).

The pathologic features of CFP are well established. These include a nonencapsulated, densely hyalinized, fibrotic proliferation with slight infiltration of lymphocytes and plasma cells as well as two types of calcification. Patients are alvvays young (mean age 17.6 years), more often they are female, and discovery of the lesion is most often fortuitous. CFP has been reported to cccur in the soft tissues of the extremities, trunk, scrotum, groin, neck or axilla and in the pleura. Some auîhors believe this proliferation to be fundamentally the result of a reactive process rather than one of a neopiastic nature and there is a possible relationship betvveen this lesion and other reported pseudotumors (4,5). Yet the pathogenesis of CFP remains uncertain. But it is perhaps a late stage of inflammatory myofibroblastic tumour. Surgical excision appears to be adequate treatment, but the number of the reported cases in the literatüre is unsatisfactory to determine therapeutic procedures (4-6).

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