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Isolated case

Primary clear cell sarcoma of tendons and aponeuroses of lymph node

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RESUMEN

El sarcoma de células claras es un tumor poco frecuente que afecta preferentemente a las extremidades. Presentamos el caso de una niña de nueve años con una masa en la rodilla que ha ido creciendo lentamente durante 14 meses. Histológicamente, se observaban restos de tejido ganglionar linfático completamente infiltrado por células poligonales o redondas, de citopasma claro y núcleo vesicular con nucléolo basófilo prominente. Las técnicas de inmunohistoquímica demostraron positividad intensa para vimentina y S100 y moderada para EMA. El examen ultraestructural mostró melanosomas aberrantes. Las características clinicopatológicas del caso apuntan con fuerza a un sarcoma de células claras primario de ganglio linfático primitivo. La localización primitiva en ganglio linfático de este raro tumor no ha sido previamente descrita en la literatura. **Rev Esp Patol 2001; 34(3): 243-248.**

Palabras clave: Sarcoma de células claras - Ganglio linfático - Primario - Melanoma de partes blandas

SUMMARY

Clear cell sarcoma is a rare tumor commonly affecting the extremities. We report on a case which originated in the lymph node. A 9-year-old girl presented with a 14-month history of a slowly growing mass at the knee. Histologically, ganglionic lymphatic tissue was found to be completely infiltrated by polygonal or round cells with clear or granular cytoplasm and vesicular nuclei with a prominent basophilic nucleolus. Immunohistochemical staining for vimentin and S-100 protein was strongly positive, and EMA was moderately positive. Ultrastructural examination showed aberrant melanosomes. The clinicopathological findings in our case strongly suggested that the lesion was primary clear cell sarcoma of lymph node. To the best of our knowledge, this is the first case presented in lymph node. **Rev Esp Patol 2001; 34(3): 243-248.**

Key words: Clear cell sarcoma - Lymph node - Primary - Malignant melanoma of soft parts

INTRODUCTION

Clear cell sarcoma of tendons and aponeuroses (CCSTA, malignant melanoma of soft parts) is a rare soft tissue malignancy that occurs most commonly in the extremities of young adults, but it has been described in several other sites, including the bone and penis (1, 2). The mean age of appearance is 27 years, but cases have

been reported in patients ranging from 7 to 83 years (3). Although lymph node metastasis is not uncommon in malignant melanoma and CCSTA (4, 5), to our knowledge, no cases of primary lymph node CCSTA have been reported. We present a case of CCSTA arising in the lymph node of a 9-year-old girl.

CASE REPORT

Clinical history

A 9-year-old girl with a 14-month history of a slowly growing mass at her left knee consulted her local physician. The tumor, which was 3 cm in diameter, was treated by local excision. Adjuvant therapy was not performed and 12 months after the diagnosis the patient is free of disease.

Materials and methods

Specimens were fixed in 10% buffered formalin for examination by light microscopy. Paraffin-embedded tissues were cut into 5 m thick sections and stained with hematoxylin-eosin. Additional stains, including periodic acid-Schiff (PAS) with and without diastase treatment, Masson-Fontana and silver impregnation for reticulin were also performed.

Immunohistochemical stainings for S-100 protein (1:2000; Dako A/S, Denmark), HMB-45 (melanoma specific antigen, 1:20; Dako A/S, Denmark), vimentin (1:200; Concepta, Barcelona, Spain), desmin (1:20 Dako A/S, Denmark), actin (1:100; Biogenex, San Ramon, CA, USA), EMA (1:100; Dako A/S, Denmark), CD31 (1:10; Dako A/S, Denmark), CD34 (1:40; Dako A/S, Denmark), FVIII (1:40; Dako A/S, Denmark), and keratins (AE1-AE3, 1:200; Signet, USA and CAM5.2, prediluted, Becton-Dickenson, USA) were performed using the avidin-biotin-peroxidase complex technique.

For electron microscopy, formalin-fixed material was post-fixed in 2.5% glutaraldehyde.

Pathological findings

The tumor, measuring 3×2 cm, was located at the external side of the left knee. The cut surface showed a white tumor, encapsulated and friably soft in consistency.

Light microscopy showed a lymph node with architectural effacement by a neoplasm, growing in nests surrounded by collagenous septa. The neoplastic cells were polygonal or round with vesicular nuclei and prominent basophilic nucleoli (Fig. 1). The PAS stain showed glycogen within the slightly eosinophilic to clear cytoplasm. No multinuclear giant cells were present. The mitotic rate was low, at one or two per 10 high-power fields.

Immunohistochemical findings

Immunohistochemical staining for vimentin and S-100 protein was strongly positive (Fig. 2), and there was moderate positivity for EMA. Staining for HMB-45, keratin, actin, desmin and vascular markers was negative.

Electron microscopic findings

Ultrastructurally, the tumor cells had irregular cytoplasmic borders and central nuclei. The nuclei were irregular in outline and showed pseudonuclear inclusions and prominent nucleoli. The cytoplasm contained swollen mitochondria, many polyribosomes, and aggregates of rough endoplasmic reticulum. In a small number of cells, non-membrane-delineated structures were identified, most of which showed dense pigmentation and some laminations. These features are consistent with aberrant melanosomes (Fig. 3).

DISCUSSION

Clear cell sarcoma of tendons and aponeuroses was first described by Enzinger in 1965 (6) and is a malignant tumor usually arising in tendons and aponeuroses in young patients. It is associated with an unfavorable prognosis, with many patients developing recurrences and metastases in an average time from diagnosis of 2.6 and 3.5 years, respectively (3, 5). The cell of origin is thought to be derived from the neural crest, as suggested by its histopathologic, immunocytochemical, and ultrastructural similarities with another neural crest-derived tumors, namely cutaneous melanoma. However, these two entities differ cytogenetically, since a t(12;22) has only been described in CCSTA (7).

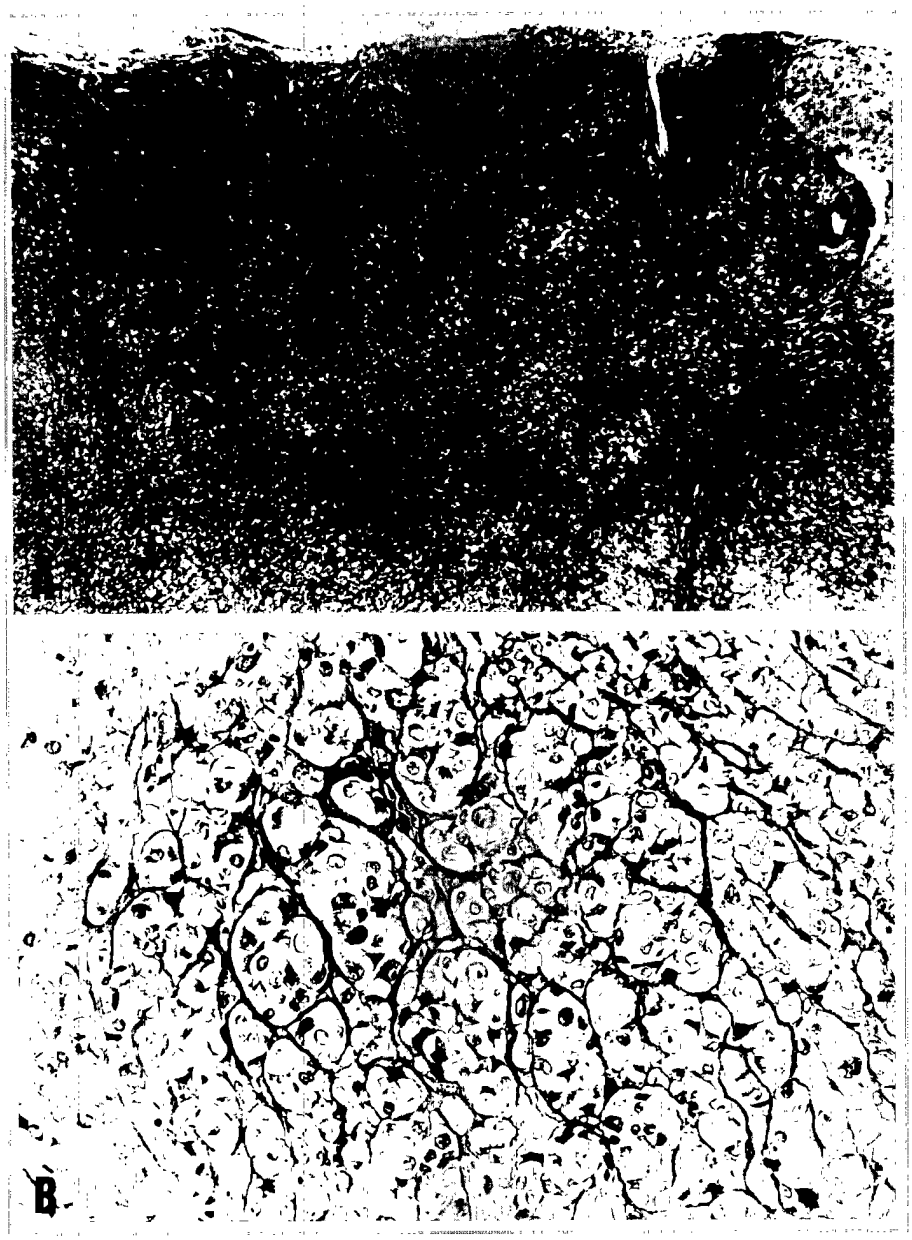

Figure 1. Histologic appearance of the tumor. (A) Lymph node affected by clear cell sarcoma of tendons and aponeuroses. A lymphoid follicle and part of the capsule of the lymph node are observed (original, HE ×100). (B) The tumor is separated into vague nests by collagenous septa. The tumor cells are characterized by clear or granular cytoplasm and vesicular nuclei containing prominent basophilic nucleoli (original, HE ×200).

Although the present case was entirely composed of round to polygonal cells, typical CCSTA may have both plump, spindled tumor cells and round to polygonal cells (8, 9). The tumor cells also show an epithelioid appearance. Delicate fibrous septa usually separate tumor nests. As in this case, mitotic index is usually low.

Immunohistochemical studies revealed that the tumor cells were positive for vimentin and S-100 protein, as usually occurs in CCSTA. In contrast, HMB-45 immunoreactivity was not detected, but some HMB-45-negative cases have been reported. Immunoreactivity for EMA has also been described in CCSTA (10, 11). At the ultrastructural level, the presence of premelanosomes was observed, consistent with previous reports (3, 8, 12).

These morphologic and immunohistochemical findings coincide with malignant melanoma. Therefore, the

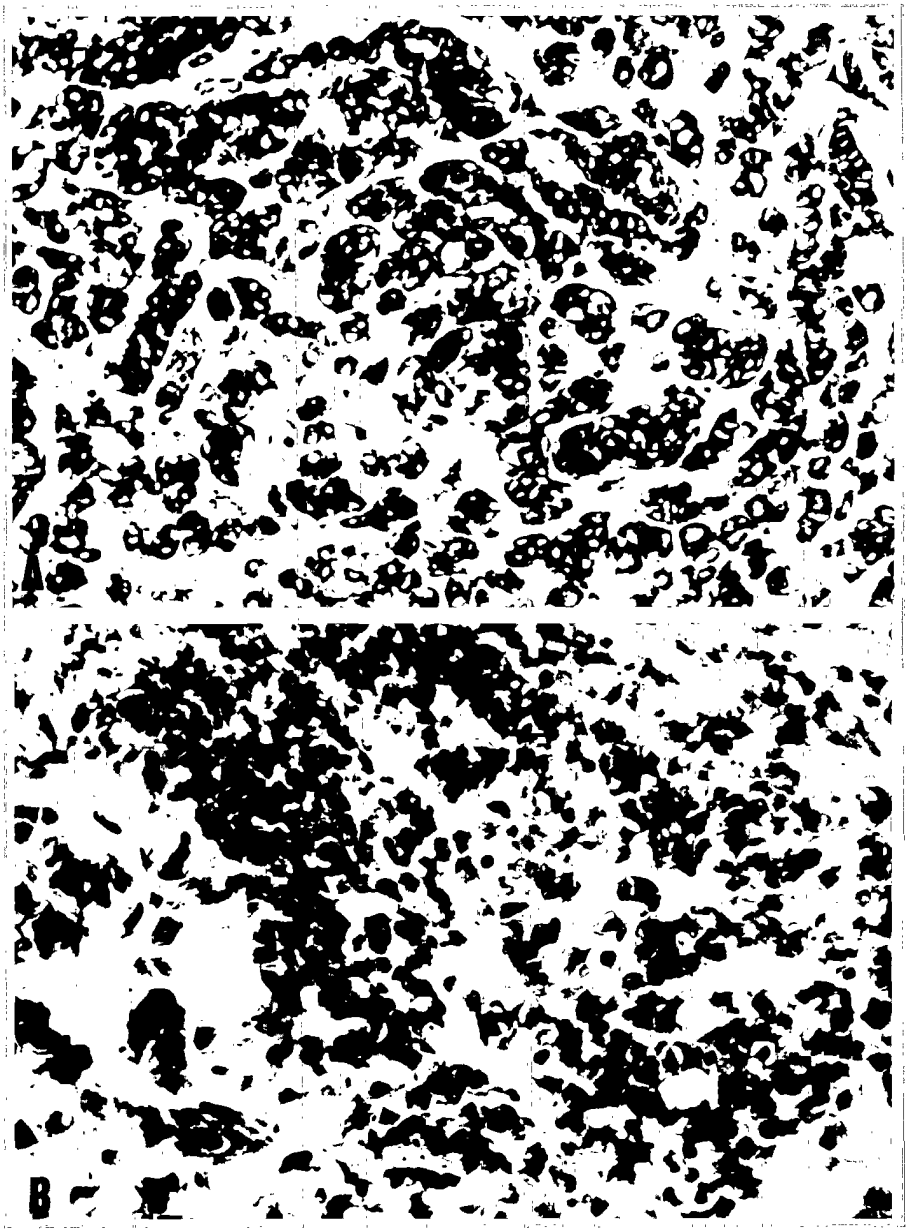

Figure 2. Immunohistochemistry of the tumor cells. The cells show strongly positive immunoreactivity for vimentin (A) and S-100 protein (B) (original, ×200).

cytogenetics, RT-PCR or FISH studies demonstrating a t(12;12) would be of great diagnostic value in our case, as it is presumed to be a specific genetic marker for CCSTA (13, 14). Unfortunately, in our case fresh or frozen tissue was not available to reliably perform genetic studies for this translocation. However, there were some clinicopathological features that made the possibility of a malignant melanoma very unlikely, such as the age of the patient and the low mitotic index.

It is not uncommon for CCSTA to metastasize to lymph node (5, 9, 15). Similarly, malignant melanoma metastatic to lymph node from an unknown primary site has been well documented (16, 17). It has been suggested that some of these cases could involve true primary melanoma of the lymph node arising in subcapsular nevus cell aggregates (18, 19). Radiological studies, including computed tomography and magnetic resonance, as well as all other diagnostic investigations

Figure 3. Ultrastructural appearance of clear cell sarcoma of tendons and aponeuroses showing tumor cell with irregular nuclear profiles, and numerous aberrant melanosomes (inset) (original, ×5,400; inset, ×45.000).

revealed no possible primary site other than the lymph node of the knee. In addition, the patient remains disease-free 12 months after surgery.

Although another unknown primary site cannot be completely excluded, the clinicopathological findings in our case strongly suggested that the lesion was a primary CCSTA of lymph node. We suggest this is another diagnostic possibility to be kept in mind when assessing a lymph node tumor resembling malignant melanoma in a young patient.

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