Soft-tissue multifocal fibromatosis and bone dysplasia in the lower limbs: exceptional association in three children

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Learning objectives

- To present three children with multicentric musculoaponeurotic aggressive fibromatosis and skeletal dysplasia associated of lower extremity.
- Identify the main imaging findings and the unfavorable developments of this rare association.
- To establish the hypothesis of a single mesenchymal congenital disorder that causes both conditions.
Background

Musculoaponeurotic aggressive fibromatosis (FMA) is a subtype of fibromatosis of intermediate malignancy, with a tendency to local invasion and recurrence but not metastasize. Its exceptional partnership with skeletal dysplasia its multiplicity increase the complexity of management.

We present three cases of boys of 1, 8 and 10 years old. In all three cases there was more than one focus of FMA in different locations of the lower extremity and fibula bone dysplastic changes (curvature, cortical thinning and widening medullary), and sometimes tibia or foot bones were involved.

The first case is a child of 8 years with two soft tissue tumors, one in his left buttock and the other in the 2nd left toe which were extirpated at another hospital diagnosed as fibrosarcoma. He is submitted by recurrence of both lesions (Figures 1 and 2). Extrinsic bone deformation is seen in the 2nd finger because adjoining fibromatosis and first grade dysplastic changes (Fig. 3) as well as in the tibia and fibula the same side (pictures 4 and 5). The resection of recurrences confirmed the diagnosis of FMA. Two years later appears a new mass in ipsilateral popliteal fossa. The masses successive recur after excision (Fig. 6), compelling the progressive amputation of the lower extremity, up to hip disarticulation seven years following initial diagnosis (Figure 7).

The second case concerns a child from birth controlled for enlargement and left fibular curvature (Fig. 8), which is treated with corrective osteotomy at 18 months. The histological study of the excised fragment showed no abnormalities. During the postoperative period appears a mass of scar which it grows progressively during two years (Figure 9), resecting with pathologic diagnosis of FMA. Nine months later the excised tumor relapse and another lesion appears next shallower (Figure 10). Adjuvant chemotherapy was decided remaining stables both lesions and postoperative deformity of the fibula to the present, 12 years after the debut (Image 11).

The third case is a 10 year old boy which has three soft tissue masses on the left foot on the back of the 3rd finger (Figure 12), on the back of the midfoot and space preaquíleo (Picture 13). Associates extrinsic bony changes in the proximal phalanx of the third finger (Figure 14) and enlargement dysplastic of the fibula (Figure 15). The masses relapsed within 2 years after surgery, and new nodules appear in popliteal fossa (Picture 16). Chemotherapy was indicated and nowadays stability was noted, four years after the debut.

The radiological manifestations of the FMA are well known (Robbin MR and cols; Imaging of Musculoskeletal Fibromatosis. Radiographics 2001, 21:585-600). However, the multicentric FMA in association with skeletal dysplasia is very rare. We found only
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Conclusion

- Musculoaponeurotic fibromatosis Aggressive (FMA) is a rare tumour that is exceptionally associated with bone dysplasia. In these cases, the FMA use to be multicentric, often asynchronous and relapsing.
- The three cases presented had affection of left leg and all have fibular dysplasia, which appears incurved and widened, which could be a key to this partnership.
- Neoplasia and dysplasia may have a common origin, which would be reinforced by the young age of our patients’ debut.
References

Aggressive fibromatosis in children and adolescents: the Italian experience.

