A case of congenital fibrosarcoma
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Section: Musculoskeletal system
Case Type: Clinical Cases
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Patient: 3 months, female

Clinical History:
A 3-month-old child with a large left leg tumefaction.

Imaging Findings:
A 3-month-old child was brought to our department with a large left leg tumefaction. A Magnetic Resonance examination (MR) was performed, showing a dishomogeneous huge mass (70mm) with liquid and necrotic areas in both spin echo T1-weighted and T2-weighted (Fig. 1). Incisional biopsy and histological study of the lesion were performed and congenital fibrosarcoma diagnosis was done. A similar smaller mass appeared at the posterior aspect of the right hemithorax within the following weeks.
Chemotherapy with Vincristine and Actinomycin-D was initiated. Chest and abdominal Computer Tomography (CT) was performed for staging. It showed additional lesions at right trapezius muscle and at the right piriformis muscle (Fig. 2). A MR examination was performed, confirming the presence of all these lesions with necrotic and liquid characteristics (Fig. 3).
Subsequently, a new MR study was performed to evaluate chemotherapy response of the disease. It showed a significant decrease of the mass lesions at the trapezius and piriformis muscles and of the leg lesion (Fig. 4).
Surgical treatment is planned after further chemotherapy.

Discussion:
Fibrosarcoma accounts for 9,5% of congenital neoplasias and for 3% of childhood tumours. It is called “congenital” because it is present at birth in approximately 37% of the cases, as a swelling or a mass that usually affects the distal portion of the extremities. Location at the trunk, head and neck regions is also possible. Usually, the mass becomes evident during the first year of life.
Congenital fibrosarcoma is considered a separate entity to its adult counterpart because of its clinical course is more favorable and metastatic spread is rare. Furthermore, congenital forms are associated the chromosomal translocation t(12;15); (p13;q25) with ETV6-NTRK3 gene fusion.
Differential diagnosis includes rhabdomyosarcoma, vascular tumours and infantile myofibromatosis. Histological analysis is mandatory, because clinical and imaging aspects are non-specific.
The most common radiological finding is a fast growing mass in the soft tissues. Radiographs can show bone incurvation, cortical swelling and rarely destruction of adjacent bones. Ultrasound (US) evaluation is useful in assessing the cystic,solid or vascular nature of the lesions. Computer Tomography (CT) examination may assess tumor size and bone involvement. On MRI, spin echo T1-weighted images before and after gadolinium and T2-weighted images are sufficient to characterize the lesions; fat suppression images add additional information if the tumour displays heterogeneous components. Scintigraphy with 99mTc-metilendifosfonato may show activity of adjacent bones and the presence of bone metastases.
Despite rapid growth and a high degree of cellularity, treatment with wide local excision without additional radiation-
or chemotherapy is usually successful in most cases. In some cases amputation is necessary if the extent or large size of the tumour precludes surgical therapy. Chemotherapy is used in non operable tumours and as neoadjuvant therapy is useful for decreasing tumour bulk. Evidences suggest that the ETV6-NTRK3 gene fusion may underlie the distinctive biological properties of these tumours and may also indicate tumour chemosensitivity. Congenital fibrosarcoma has a relatively good prognosis and only rarely metastasises. Prognosis is better if the chromosomal translocation is found. Recurrence rate is 17-40% with a five years survival of 84-93%. Metastasis rate depends on the site of the primitive tumour: 8% for location at the limbs, 26% for axial location. One case of spontaneous regression of neonatal fibrosarcoma is described in literature.

**Differential Diagnosis List:** Multisegmental neonatal fibrosarcoma.

**Final Diagnosis:** Multisegmental neonatal fibrosarcoma.

**References:**


Description: On T2-weighted a heterogeneous huge mass (70mm) with liquid and necrotic areas was demonstrated. Origin:
**Description:** On T2-weighted a heterogeneous huge mass (70mm) with liquid and necrotic areas was demonstrated. **Origin:**
Description: CT examination showed a tumefaction at the right piriformis muscle (20mm) and... Origin:
Description: ...the presence of mass at the right trapezius muscle(14mm). Origin:
Figure 3

Description: MR examination shows left leg lesion... Origin:
Description: ...with necrotic and liquid characteristics on T2-weighted images. Origin:

Description: A similar mass was seen at the right piriformis muscle tumefaction and... Origin:
Description: ... at the right trapezius muscle. Origin:
Figure 4

Description: MR examination shows a significant decrease of leg mass (45mm). ... Origin:

Description: ... of piriformis muscles masses, and... Origin:
Description: ...of trapeze lesions. Origin: