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CASE REPORT

# Proliferative myositis in a child

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**Abstract** A case of proliferative myositis in the lumbar paraspinal muscles in a 14-year-old boy is presented. Imaging investigations including plain radiograph, ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), bone scan and positron emission tomography (PET) were suggestive of an inflammatory process such as myositis ossificans. The diagnosis was made by incisional biopsy. More pronounced edema, more muscle fiber necrosis and a higher cellularity were found compared to adult cases. The karyotype of the lesion was normal. Clinically, the mass disappeared spontaneously. After 24 months, asymptomatic bridging ossification between the third and fourth lumbar vertebrae was noted.

**Key words** Proliferative myositis · Childhood · Lumbar muscles · Ossification · X-ray · CT · MRI · PET

## Introduction

Proliferative myositis (PM) was first described by Kern [1] in 1960, and is an uncommon but distinctive, self-limiting intramuscular pseudosarcomatous inflammatory process. It presents clinically as a rapidly growing mass at a median age of 50 years. We report a case of PM in a child, diagnosed by incisional biopsy and treated conservatively.

## Case report

A 14-year-old Caucasian boy presented with a painful left dorsolumbar paraspinal mass. Three weeks earlier, he had sustained a blunt injury to the sternum. The day following the injury, he had experienced increasing pain in the left gluteal region. He had received five physiotherapy sessions and local warmth had been applied. One week later, the pain extended to the left dorsolumbar paraspinal region, whereupon he developed an antalgic scoliosis. Another week later, the pain radiated to the left groin and hip. At this time, mild pyrexia (37.5°C) had been noted. He had no past medical history apart from mild asthma.

Physical examination revealed an extreme antalgic scoliosis. The left dorsolumbar paraspinal region was very tender on palpation and moder-

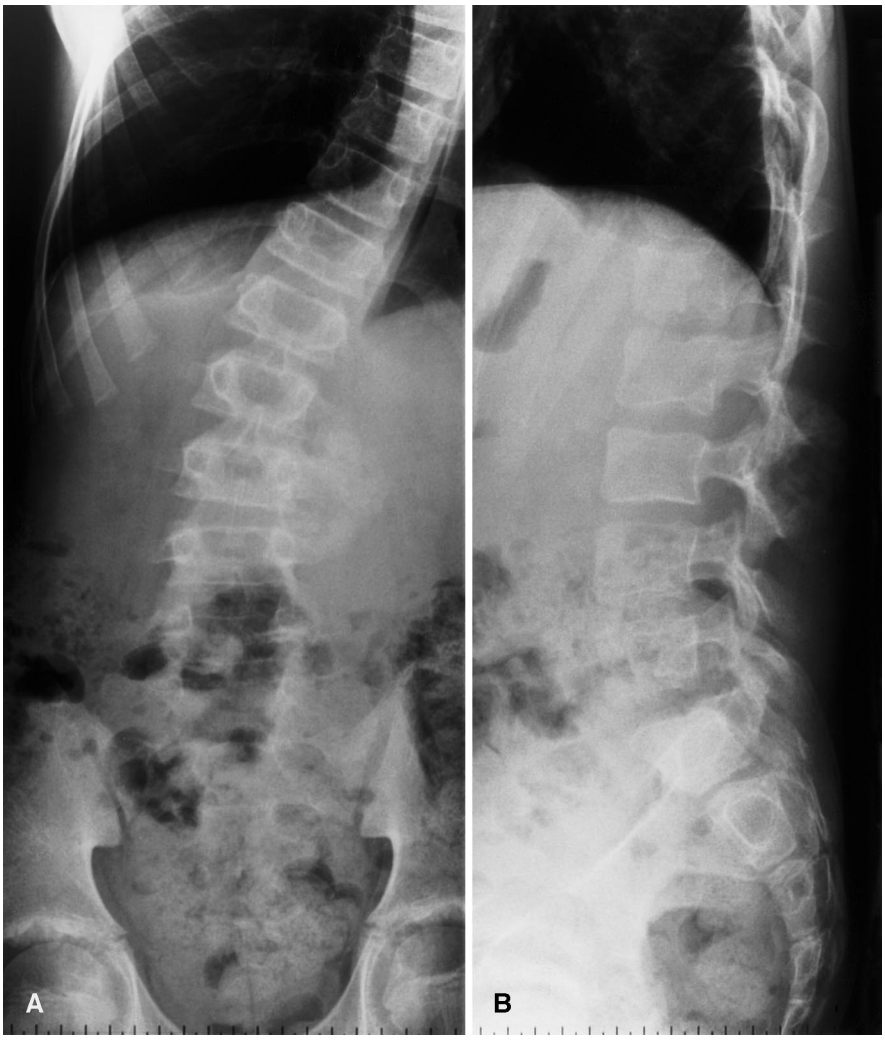



**Fig. 1A,B** Radiographs of the lumbar vertebral column show a confined area of calcification on the left side of L3 and posterior

ately swollen. There was no skin discoloration nor local warmth. No other abnormalities were noted on physical examination.

The erythrocyte sedimentation rate (ESR) was 31 mm in 1 h, and C-reactive protein as well as leukocyte count were normal. Tumor markers carcino embryonal antigen (CEA), alpha-fetoprotein (αFP), beta human chorionic gonadotropin (βHCG) and neuron-specific enolase (NSE) were normal.

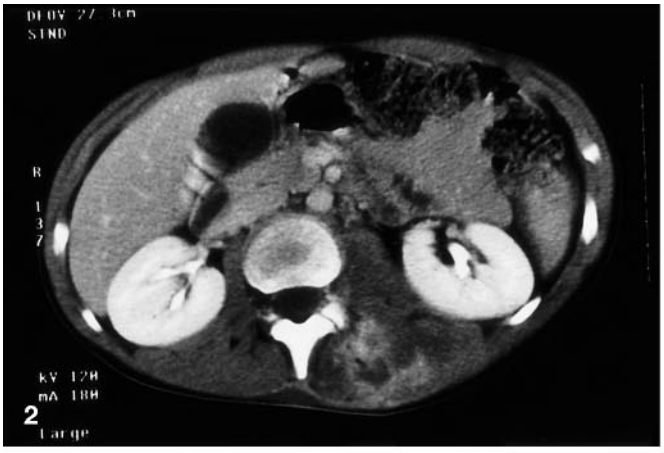
A radiograph of the lumbar vertebral column showed a confined area of homogeneous calcification on the left side of L3 (Fig. 1). Ultrasound showed an inhomogeneous mass with calcifications. CT scan (Fig. 2) revealed a heterogeneous hypodense mass with contrast enhancement. On T1-weighted MR images (Fig. 3), a markedly contrast-enhanced nodular lesion of 4 cm in the left sacrospinalis muscle was found, surrounded by a moderately contrast-enhanced inhomogeneous infiltration of the quadratus lumborum muscle as well as of the left psoas muscle.

On the MDP (methylene diphosphonate) bone scan a focal area of intense radiotracer accumulation was seen adjacent to the left part of the third lumbar vertebra. A second focal area of radiotracer accumulation was seen on the dorsal part of the seventh rib, and was attributed to an old fracture. A whole-body FDG (fluorine-18 deoxyglucose) PET (positron emission tomography) scan showed an intense hot spot lateral to the lumbar spine (Fig. 4A). Clinical, radiological and scintigraphic findings suggested an inflammatory process such as myositis ossificans. An incisional biopsy (2 $cm^3$) of the nodular lesion at the edge of the dorsolumbar spine was performed 3 weeks after the onset of the pain. Macroscopically, the lesion was gray and friable, with no distinct edge to the surrounding pale and edematous muscle fibers.

Histological examination (Fig. 5) revealed a proliferation composed of two cell populations: a minor population of small spindle cells with an elongated nucleus and a major population of ganglion-like giant cells with a large nucleus, a prominent nucleolus, and a fair amount of basophilic cytoplasm. No obvious cellular pleomorphism was present. Mitoses were frequent but not atypical. A typical checker-board pattern was formed by the proliferation of cells with marked edema between the perimysial septa. Focal necrosis of muscle fibers at the periphery of the lesion was seen. Few lymphocytes were seen in the lesion. Polymorphonuclear cells were notably absent. There was no evidence of cartilage or osteoid formation.

Immunohistochemical analysis revealed vimentin and alpha smooth muscle actin by the ganglion-like cells. These cells did not express desmin or myosin. Because of the typical morphological and immunophenotypic findings, the diagnosis of proliferative myositis (PM) was made. Cytogenetic analysis, after short-term culture of part of the specimen, revealed a normal chromosomal pattern46, XY.

Treatment consisted of rest and analgesics. Ibuprofen 200 mg qid was more effective than paracetamol



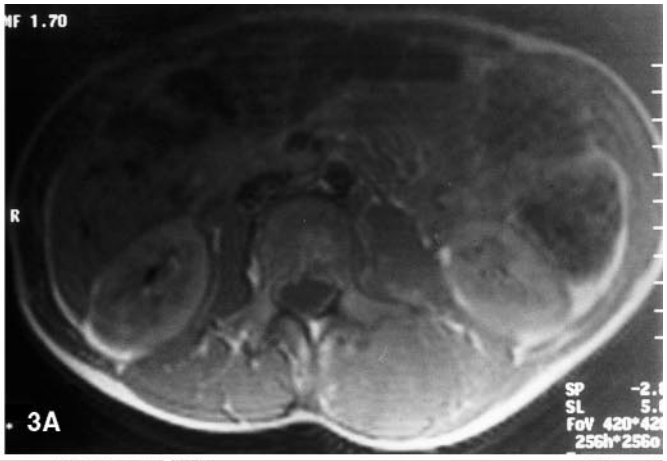
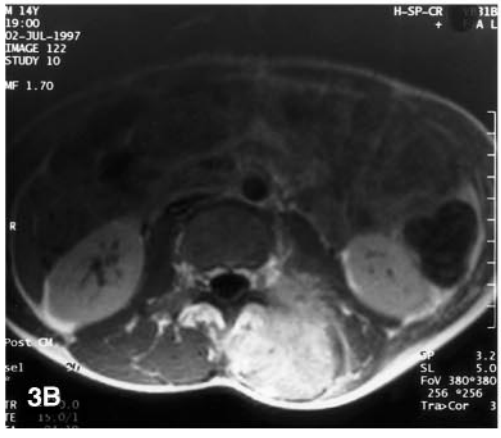




**Fig. 2** Computed tomographic (CT) scan shows an inhomogeneous hypodense mass with contrast enhancement



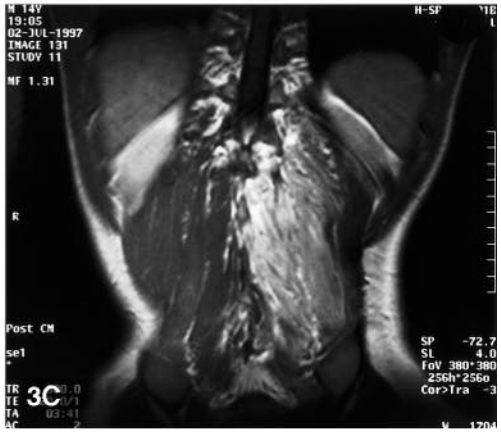
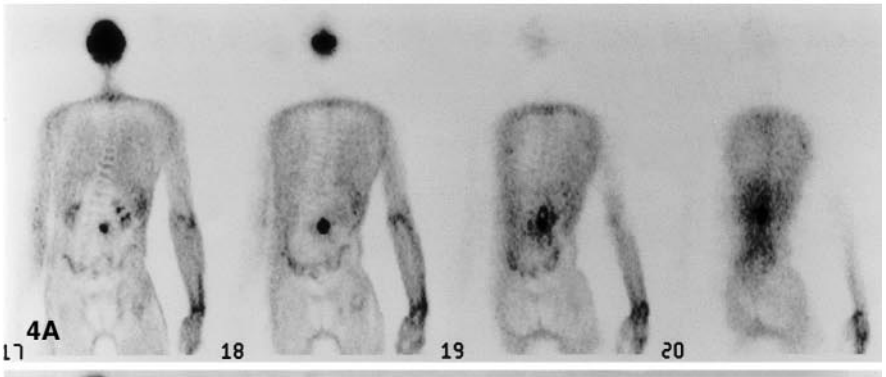




**Fig. 3** **A** Pre-contrast T1-weighted axial magnetic resonance (MR) image shows a homogeneous hypointense mass in the left sacrospinal muscle. **B** Gd-enhanced T1-weighted axial MR image shows a contrast-enhanced nodular lesion in the left sacrospinal muscle surrounded by an inhomogeneous infiltration of the quadratus lumborum muscle and the psoas muscle. **C** Gd-enhanced T1-weighted coronal MR image shows enhancement of the entire left quadratus lumborum muscle



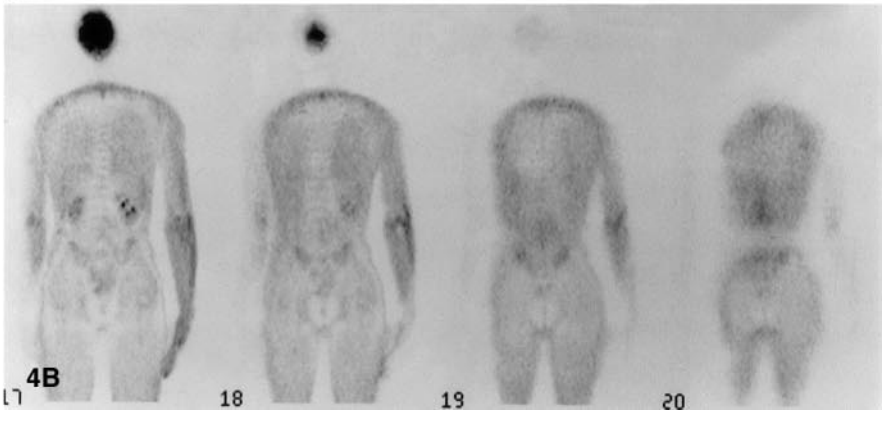




**Fig. 4** **A** Initial whole-body fluorine-18 deoxyglucose positron emission tomographic (PET) scan shows an intense focus of activity in the left paraspinal muscles. **B** Three months after the onset of the pain, the paraspinal focus of activity has disappeared on a whole-body FDG PET scan

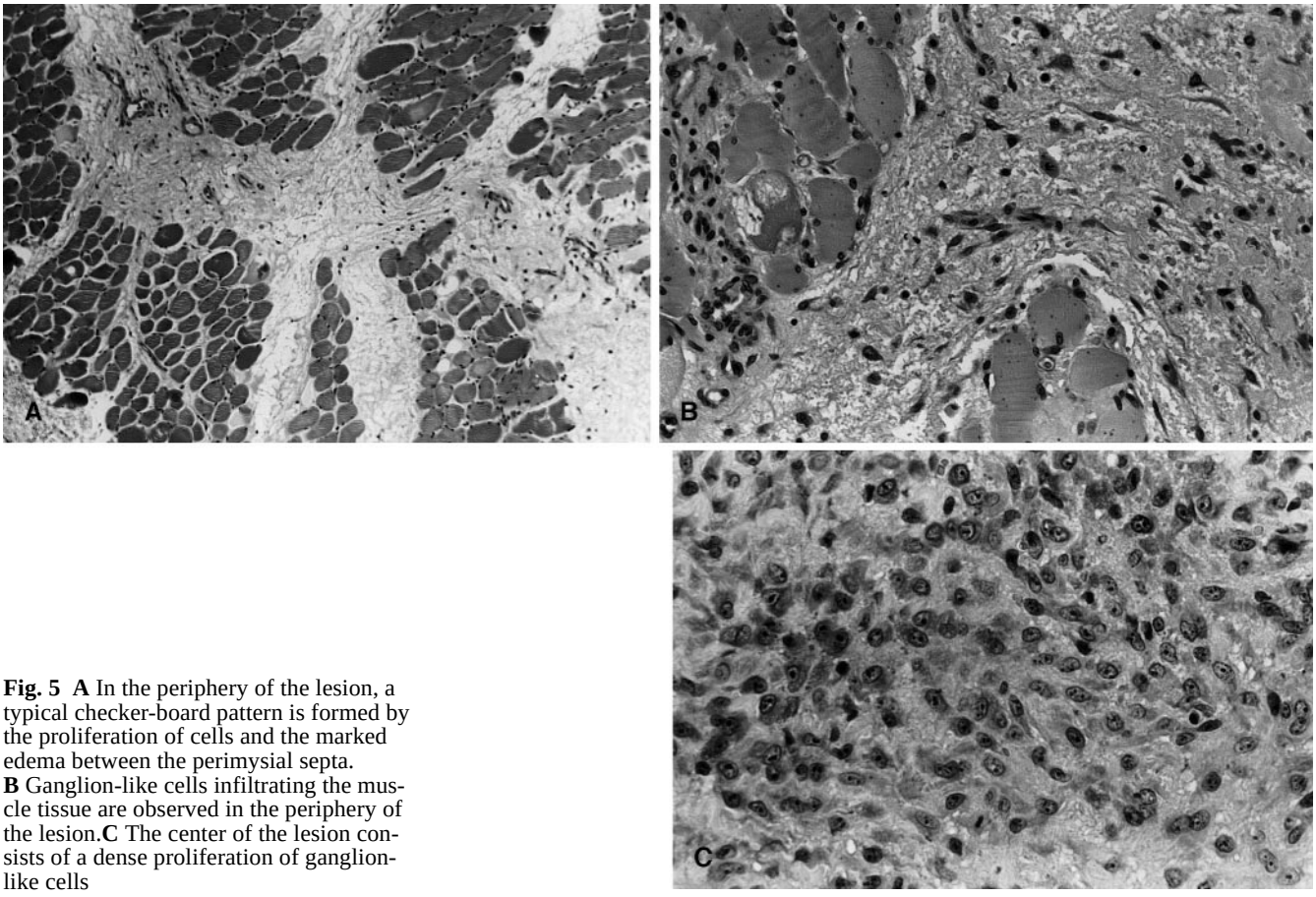
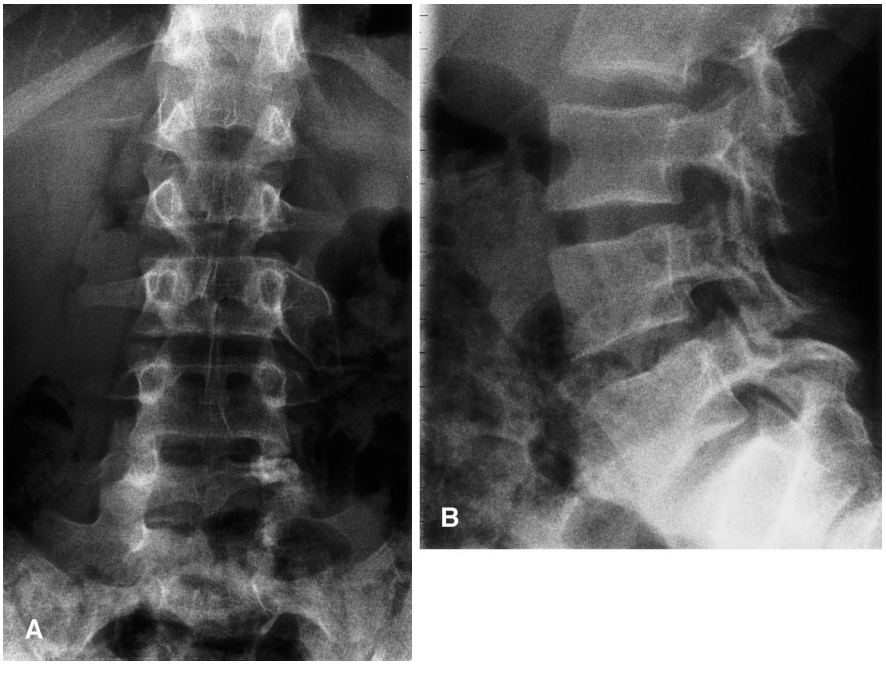



**Fig. 5** **A** In the periphery of the lesion, a typical checker-board pattern is formed by the proliferation of cells and the marked edema between the perimysial septa. **B** Ganglion-like cells infiltrating the muscle tissue are observed in the periphery of the lesion.**C** The center of the lesion consists of a dense proliferation of ganglion-like cells

**Fig. 6A,B** Radiographs of the lumbar vertebral column after 24 months show bridging ossification between the transverse and spinous processes of the third and fourth lumbar vertebrae




500 mg qid. Physiotherapy and exercise were discouraged.

Seven weeks after the onset of the pain, the patient was seen in the outpatient clinic. Pain control was good, the antalgic scoliosis had almost disappeared, and the local swelling had decreased, although there was still some induration under the scar. Three months after the onset of the pain, the swelling was gone and pain was only occasionally felt on physical exercise.

A new PET scan showed that the paraspinal area of activity had disappeared (Fig. 4B).

**Table 1** Proliferative myositis in children: histological features

| Case | Reference | Color | Demarcation | Cellularity | Mitoses | Neutrophils | Edema | Necrosis | Spindle cells |
|---|---|---|---|---|---|---|---|---|---|
| 1 | 24 | Gray | Poor | High | Occasional | Absent | Present | Absent | Present |
| 2 | 25 | Gray-white | Good | Variable | Occasional | Absent | Variable | Present | Variable |
| 3 | 26 | Gray-brown | Poor | High | Absent | Present | Present | Present | Present |
| 4 | 4 | Gray | Poor | High | Occasional | Absent | Marked | Present | Present |
| 5 | 13 | White | Poor | High | Absent | Absent | Present | Absent | Present |
| 6 | 3 | White | Poor | High | Absent | Present | Marked | Absent | Present |
| 7 | 14 | White | Poor | High | Absent | Absent | Present | Absent | Absent |
| 8 | This report | Gray | Poor | High | Frequent | Absent | Marked | Marked | Scare |

Twenty-four months after the onset of the pain, the patient was asymptomatic. No mass was felt and the mobility of the spine was normal. An X-ray of the lumbar vertebral column, however, showed bridging ossification between the transverse and spinous processes of the third and fourth lumbar vertebrae (Fig. 6).

## Discussion

Proliferative myositis is a rare benign pseudosarcomatous lesion. Accurate knowledge and diagnosis of PM is important, as these lesions can clinically be mistaken for a malignant tumor, which could result in unnecessary radical surgery. We are aware of only seven reported cases in children in the world literature [2–8]. PM appears as a very rapidly growing mass that is often painful [9]. The etiology of PM is unknown, although recent trauma is noted in one-third of cases [9].

In our case, amorphous calcification matured into bone. The absence of osteoid in the biopsy was probably due to a sampling error. Up to 10% of cases of PM contain small foci of metaplastic bone or cartilage [10]. Radiological differential diagnosis of amorphous calcification in soft tissue tumors includes nodular fasciitis, which can present with subcutaneous and fascial calcification; myositis ossificans, in which a characteristic peripheral shell is formed with maturation; and synovial sarcoma, which presents with calcification in up to 30% of cases [11].

Radiological differential diagnosis of bone formation in soft tissue lesions includes malignant tumors such as extraskeletal osteogenic sarcoma; dedifferentiated liposarcoma; and benign tumors such as ossifying lipoma, chondroma and hemangioma [11].

In two cases, including ours, results of ultrasonography were reported [12]. Whereas in our case the inhomogeneous structure and the calcifications were the predominant features, Sarteschi et al. described a characteristic “scaffolding” between continuous muscle bundles on the longitudinal scan and a “checkerboard pattern” on the transverse scan, corresponding to the typical histologic architecture of proliferative myositis [12].

In eight cases, including ours, detailed results of CT scan imaging were reported [3, 9, 13–17]. A characteristic CT pattern was not identified except for poor demarcation in all but one case. Without contrast, the mass was hypodense or isodense to the surrounding muscle. Contrast enhancement was homogeneous, heterogeneous, or absent. A hypodense rim was observed in one case. In three cases, including ours, the results of MRI imaging were reported [4, 5]. MRI showed an ill-defined mass that was hyperintense on T2-weighted images and hypointense on T1-weighted images. In two cases, an area of perilesional edema was found. Contrast, reported in our case only, showed enhancement of the lesion. The ill-defined infiltration in the surrounding muscles was suggestive of an inflammatory process.

MDP uptake on bone scan in proliferative myositis has not been described before, but can be explained by uptake of MDP in extraosseous calcifications [18].

FDG accumulation is not specific for malignancy, but occurs in all lesions with increased glucose metabolism. Therefore, FDG PET does not allow differentiation between malignant tumors and focal inflammation [19].

An excised lesion of PM appears macroscopically as a poorly demarcated gray-white scar-like lesion infiltrating the surrounding muscles. On microscopic examination, three concentric areas can be identified [20]. In the periphery, the perimysium and endomysium are infiltrated by a loose tissue of elongated spindle-shaped cells, forming a characteristic checker-board pattern. The muscle fibers are generally uninvolved. In the intermediate area, giant ganglion-cell-like cells are admixed with the spindle-shaped cells; in both, mitotic figures are common. The giant cells show an abundant, deeply staining basophilic cytoplasm and have one or two eccentrically placed vesicular nuclei with prominent nucleoli. The central area of the lesion predominantly shows giant cells in a delicate network of collagenous fibers, which has replaced the muscle tissue.

Immunohistochemically, the giant cells stain positive for vimentin and smooth muscle actin, and rarely for desmin and myosin. They do not stain for myoglobin. Ultrastructurally, they show the features of myofibroblasts [21, 22]. Focal calcification, as seen in our case, exceptional-

ly occurs, but is always less conspicuous than in myositis ossificans [23].

In the case reported, the architecture and the presence of ganglion-like cells without overt atypia were typical for PM. The immunophenotype of the giant cells, which resembles that of myofibroblasts but not of muscle cells or ganglion cells, allowed exclusion of rhabdomyosarcoma and ganglioneuroblastoma.

To our knowledge, PM has been reported in only eight children [2–8], including our case. The clinical features (sex, location, size, duration of symptoms, pain, history of trauma) are not different from adult cases. However, the histology of PM in children has specific characteristics (Table 1). As in the current case, a more pronounced edema, more muscle fiber necrosis, and a higher cellularity are often found compared to adult cases. Of note in the current case is the homogeneous proliferation of ganglion-like cells, with only a minimal presence of spindle cells. These features have been noted in two other pediatric cases of PM [5, 7] and contrast with adult PM, where the proliferation of ganglion-like cells and spindle cells are equally abundant. Interestingly, inflammatory cells are scarce in our case, in contrast to two other pediatric cases of PM [2, 8].

The karyotype in our case was normal. Two other cases of PM have been cytogenetically investigated, in which a trisomy 2 and a 46,xx, t(6;14)(q23;q23) were found [24, 25].

The first reported cases of PM were treated aggressively as malignant tumors with radical excision, sometimes in combination with lymphadenectomy, radiotherapy or chemotherapy [9]. After the clear descriptions by Kern [1] and Enzinger et al. [26] of the histological features and the benign inflammatory nature of PM, marginal excision became standard therapy, with no reported recurrences [9]. However, treatment by excision of a self-limiting inflammatory lesion does not seem reasonable. Recently, 11 patients have been reported, including our patient, in whom only an incisional biopsy or fine needle aspiration biopsy (FNAB) was done, without excision [3–6, 14, 17, 27, 28, 30, 31], with a follow-up of between 9 months and 11 years (median 1 year). Clinically, the lesions disappeared spontaneously in all patients. However, one patient had a recurrence after 6 months and underwent an excision, after which he remained well [14]. In our case, the lesion matured into an asymptomatic area of ossification.

In one patient, a short-term steroid course was given, with complete disappearance of a 3-cm mass within 1 week [17].

In our patient, who received non steroidal anti-inflammatory drugs (NSAIDs), recovery was complete too, but only after 3 months. The clinical impression was that the NSAIDs were effective in the relief of pain, but did not shorten the time to recovery. The apparent lack of a direct therapeutic action of NSAIDs is further supported by the observation of a patient with rheumatoid arthritis, who developed PM despite the fact that he was being treated with NSAIDs [32]. Physiotherapy was discouraged in our patient because of the possible triggering role of trauma in PM [9].

Proliferative myositis should be suspected when a mass develops very rapidly in a muscle compartment, and included in the differential diagnosis of soft tissue tumors. Although imaging studies may suggest an inflammatory process, incisional biopsy is recommended to establish the diagnosis. Specific histological characteristics can be found in children. Resection is not indicated, as the lesion heals spontaneously.

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