# Neonatal umbilical inflammatory myofibroblastic tumor

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**Inflammatory myofibroblastic tumors represent a tumor class of intermediate malignant potential predominantly seen in children and adolescents. Here is the first description of an inflammatory myofibroblastic tumor at the umbilicus of a neonate. In neonates the main sites of presentation are equally distributed between the thoracic and the abdominal region. In a third of the neonates the tumor is identified on an antenatal scan. The preferred treatment option is resection of the tumor. Spontaneous regression has been described.** ***Ann Pediatr Surg*** **13:160–162 © 2017 Annals of Pediatric Surgery.**

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## Introduction

Inflammatory myofibroblastic tumors (IMTs) are a rare entity of tumors with intermediate malignant potential [1–3]. We present a case of an umbilical IMT in a neonate and summarize the available literature as regards the presentation and management of IMTs in neonates [4–11].

## Case study

The baby girl was born at term at home in a northeastern province of Nigeria. Her birth weight was 2.7 kg; her Apgar score remains unknown. An antenatal ultrasound scan was not available because of the very limited healthcare resources in this region. At the end of October 2011, the 5-day-old girl was presented to the regional hospital with a fleshy mass protruding from the umbilical region. Her general examination was unremarkable; there was no evidence of dysmorphic features. The maximal dimensions of the lesion were $\sim 5 \times 3$ cm (Fig. 1). The tumor was removed under local anesthesia (General anesthesia was not readily available in the low-resource setting of northeastern Nigeria.). A small transverse incision to the right of the umbilicus allowed access to the abdominal part of the protruding lesion. The intra-abdominal component was smaller than expected and could easily be mobilized. There was no evidence of infiltration into nearby structures (e.g. round ligament/ductus venosus). Furthermore, we could not identify a patent urachal or omphalomesenteric duct. Postoperative recovery and wound healing were uneventful.

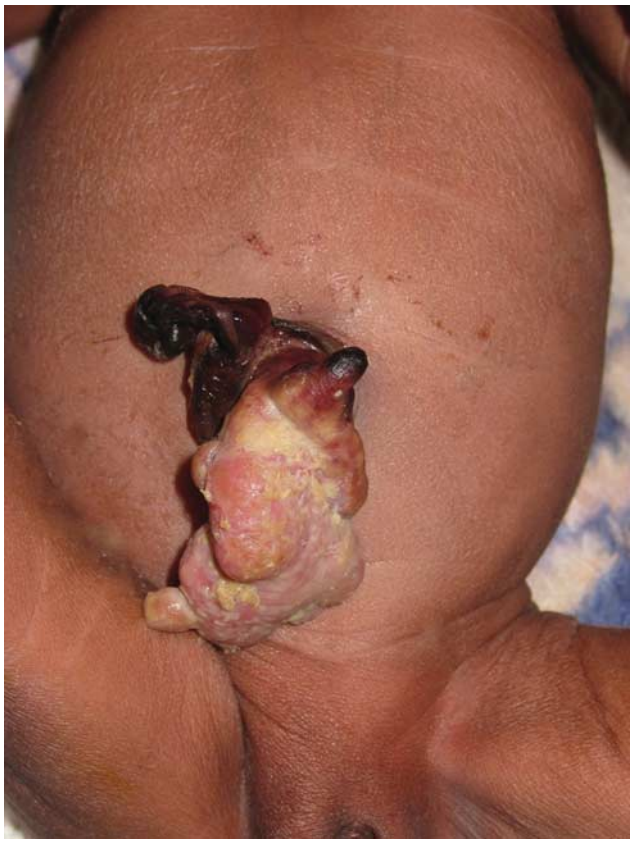
On review of the hematoxylin and eosin-stained histology slides in a German laboratory, spindle cell proliferation accompanied by a mixed inflammatory cell infiltrate was identified. The spindle cells showed increased mitoses, but no atypical mitotic figures. Distinct cellular differentiation such as cartilage or muscle striation was not found. The overall picture represents an IMT (Fig. 2). Paraffin blocks for further immunohistochemical analysis, particularly to identify expression of anaplastic lymphoma kinase (ALK), were not available.

At the first follow-up at around 4 weeks, the baby girl was well and thriving. She continued to develop appropriately at 6 months of age.

## Discussion

On review of the literature, eight more neonates with IMT in various locations were identified (Table 1). The most common sites of presentation were the thorax and

**Fig. 1**



Inflammatory myofibroblastic tumor at the umbilicus of a neonate.

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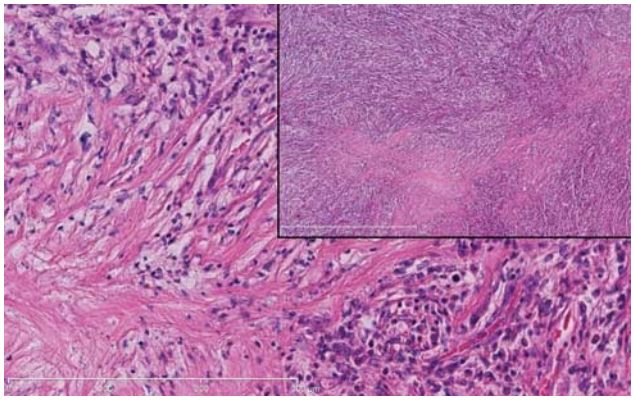
abdomen. Other anatomical regions were the brain, the parapharyngeal region, and the bladder [5,8,10]. Respiratory distress at birth is a leading symptom [4,7–9]. Antenatal identification of the tumor, or indirect signs of a lesion (e.g. hydrops), was observed in a third of the cases [6,7,9].

IMTs are a distinct entity within the group of so-called inflammatory pseudotumors [3]. IMTs are characterized by myofibroblastic spindle cells accompanied by an inflammatory infiltrate of variable degree [12]. IMT was first recognized in lung tissue by Brunn in 1939 and has since been identified in most organ systems [1,12–14]. Historically, IMTs were considered to arise as a result of an exaggerated reactive or reparative process to tissue injury [1]. However, since the identification of abnormalities on chromosome 2p23 leading to p80 and ALK expression in some IMTs, these tumors are considered neoplastic lesions with an intermediate neoplastic potential [10,15–17]. ALK expression is seen in around 50% of the tumors [2,10,15].

**Fig. 2**



Spindle cell pattern of inflammatory myofibroblastic tumor. (hematoxylin and eosin staining; main picture scale bar 300 μm; top right picture scale bar 1 mm).

There is evidence for local recurrence occurring in approximately a quarter of the affected patients; metastatic spread is rarely seen [3,17]. Overall, IMTs are most commonly seen in children and young adults, but they can occur throughout life [3,10]. Nearly two-thirds to three-quarters of the lesions are identified in the abdominopelvic region; one-fifth of the IMTs present in the thoracic cavity either affect the lungs directly or are located in the mediastinum [3,10].

The subgroup of neonates with IMT show an equal distribution between thoracic and abdominal presentation; probably a thoracic lesion is easily noticeable through symptoms such as respiratory distress compared with a relatively small abdominal mass (Table 1). Nevertheless, antenatal identification of the tumor was observed in a third of the cases [6,7,9].

The mainstay of treatment is surgical resection [17]. Castañón *et al.* [9] reported a large left-sided thoracic tumor, which was embolized before its successful surgical removal. The 'Italian Cooperative Group Studies' reported experience with chemotherapy in a small group of patients – mainly teenagers – with recurrence, although with variable response [17].

NSAIDs are being suggested as a treatment option because of their anti-inflammatory and antiangiogenic effects [18]. In some instances the use of NSAIDs resulted in tumor regression in older children, whereas others reported no response to NSAIDs [10,19]. Similarly, the anecdotal usage of steroids show mixed results: an elderly patient with a renal IMT showed a good response to steroids [20]. A neonate with a parapharngeal mass received steroids, which had no effect on the lesion; the baby died 5 weeks later of respiratory distress [8].

Spontaneous regression of the thoracic component after excision of the abdominal part of a multifocal IMT has been observed [7].

**Table 1 Inflammatory myofibroblastic tumors in neonates**

| References | Gestation/age/sex | Presentation | Location | Treatment | Follow-up |
|---|---|---|---|---|---|
| Alobeid *et al.* [4] | 35 weeks/1 day/female | Respiratory distress | Right thoracic mass | Lobectomy | 12 months (NED) |
| Asanuma *et al.* [5] | FT/7 days/female | Hematuria | Bladder | Excision | 12 months (NED) |
| Sirvent *et al.* [6] | 39 weeks/4 months/male | Antenatal scan lumbar mass | Lumbar region | Excision | NS |
| Thompson *et al.* [7] | 39 weeks/1 day/male | Antenatal scan abdominal and thoracic mass | Anterior abdominal wall and thoracic mass | Excision of abdominal tumor and regression of thoracic component | 6 years (NED) |
| Klein *et al.* [8] | 38 weeks/1 day/female | Inspiratory stridor | Pharynx | Steroids only | 5 weeks (Died) |
| Castañón *et al.* [9] | 38 weeks/1 day/male | Antenatal scan hydramnios, respiratory distress | Left thoracic mass | Embolization and excision | 20 months (NED) |
| Coffin *et al.* [10] | FT/3 weeks/male | Unknown | Brain | Excision | NS |
| Fragoso *et al.* [11] | NS/28 days/female | Palpable abdominal mass | Adrenal region | Excision of adrenal mass and kidney | 13 years (NED) |
| This study (2015) | FT/3 days/female | Visible tumor | Umbilicus | Excision | 6 months (NED) |

Age, age at operation; FT, full term; NED, no evidence of disease; NS, not specified.

### Conflicts of interest

There are no conflicts of interest.

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