

## Diagnosis and treatment of inflammatory myofibroblastoma in children and adolescents

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*To the Editor:* Inflammatory myofibroblastoma (IMT) is a rare type of intermediate tumor with malignant potential that often affects children and young adults. Although IMT can arise from various organs, most originate in the lungs, abdomen/pelvis or retroperitoneum. These tumors are thought to arise via a benign inflammatory process, although some exhibit malignant behavior or undergo a malignant transformation. There are no uniform guidelines for the clinical management of IMT.<sup>[1,2]</sup> Currently, surgery is first choice of treatment, although other therapeutic options may be considered when total excision is not possible.<sup>[2,3]</sup> In this report, we summarize the clinical data of 16 patients aged  $\leq 18$  years who were diagnosed pathologically with IMT at our department between 2013 and 2018, review the literature and share our experiences with the diagnosis and treatment of this disease.

The patients had a mean age of  $8.9 \pm 5.7$  years (range, 0.9–18.0 years) at presentation and a male to female ratio of 1:1. The mean follow-up duration was  $2.3 \pm 1.5$  years (range, 0–4.7 years), and 2 children were lost to follow-up. The following clinical manifestations of intraperitoneal IMT were observed: abdominal distension, abdominal pain, palpable mass, blood stool, fever, yellowed skin and mucous membranes, and other digestive symptoms. Pulmonary IMT was associated with the following clinical manifestations: cough, expectoration, dyspnea, and chest pain. Bladder IMT manifested as hematuria, while maxillofacial and cervical IMTs were associated with swelling and palpable mass. Additionally, seven patients presented with systemic symptoms, such as fever, anorexia, and weight loss.

Six of 16 patients presented with an elevated white blood cell count (38%), whereas 11 of 16 patients had hypochromic microcytic anemia (69%). Non-specific tumor markers were detected in 12 (75%).

In 11 cases, computed tomography (CT) revealed tumors with clear borders that were excised completely using the following procedures: lung lobectomy (left lung;  $n=1$ , right middle and lower lobe;  $n=1$ ), excision of the left superior mediastinum tumor and partial rib ( $n=1$ ), total excision of the mass with adjacent bowel ( $n=3$ ), partial cystectomy ( $n=2$ ), and total excision of the neck mass ( $n=3$ ). Nine of these 11 patients survived without recurrence, one was lost to follow-up and one died of a chest infection 1 month postoperatively.

In some cases, palliative surgical excision was selected because the tumors had invaded important adjacent tissues and organs.<sup>[3]</sup> In one case, CT revealed that a left lung IMT had squeezed and invaded the mediastinal macrovascular structure. This patient received regular chemotherapy after palliative excision and remains tumor-free. A patient with an omental IMT experienced a recurrence after palliative resection and subsequently underwent chemotherapy after resection. This patient also remains tumor free. A patient with rectal IMT experienced two postoperative recurrences that were treated surgically and remains alive but not tumor free. In another case, a liver IMT was found to have invaded all three hepatic hila [Figure 1] and was subjected to complete tumor removal and autologous orthotopic liver transplantation. A patient with pancreatic head IMT refused further treatment after a puncture biopsy and was lost to follow-up.

Most cases of pediatric IMT are benign, and the overall survival rate exceeds 90%. As noted earlier, surgical resection is the main treatment option for IMT, which can be cured if completely resected with microscopically clear margins.<sup>[3]</sup> Additionally, partial mass excision can be performed to treat locally infiltrating and anatomically complex IMTs that may not be indicated for total excision. Chemotherapy, immunoregulatory therapies, corticosteroids, radiotherapy, non-steroidal anti-inflammatory drugs, and other adjunctive therapies may also be

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**Figure 1:** Features of a liver inflammatory myofibroblastoma (IMT) on coronal computed tomography (CT) images, demonstrating invasion of the portal vein, including the left branch, with tumor embolus (A). The tumor has invaded and encircled the inferior vena cava (B). CT features of an IMT in the left lung. Transverse (C) and coronal contrast-enhanced CT images (D) demonstrate heterogeneity in the left lung tumor, which compressed and invaded the mediastinal macrovascular structure.

recommended if a complete resection is not feasible.<sup>[4]</sup> In our report, 15 of the 16 patients underwent surgical excision; of these, 12 achieved a macroscopically complete resection. Furthermore, three patients with mediastinal or abdominal vital visceral invasion achieved an incomplete resection. The residual shadow of the tumor was reduced in CT, and the effect of chemotherapy was considered. Furthermore, three patients with mediastinal or abdominal vital visceral invasion achieved an incomplete resection. Two of these patients received regular chemotherapy after palliative resection using the alternative AVCP regimen (pirarubicin + vindesine + cyclophosphamide + cis-platinum complexes) or IEV regimen (etoposide + ifosfamide + leurocristine). In these cases, chemotherapy led to reductions of the residual tumor shadow on CT images. All 16 children received anti-inflammatory therapy. Two cases of ALK-positive disease were detected by fluorescence *in situ* hybridization, and crizotinib was administered.<sup>[5]</sup> The effects of this treatment require

further study. We generally do not recommend waiting for spontaneous regression.

The IMT recurrence rates range from <2% for lung tumors to 25% for extrapulmonary lesions, and most recurrences develop within 1 year after the initial surgery. Distant metastasis is rare, occurring in <5% of cases.<sup>[1]</sup> Complete tumor excision during the initial surgery, with clear surgical margins, is the single most important factor preventing recurrence.<sup>[3]</sup> We were able to follow-up 13 of 16 cases, with a survival rate of 81%. All of these patients underwent surgical treatment. Twelve patients remained in complete remission at the last evaluation and one survived with partial remission. Two children were lost to follow-up: one had achieved a complete resection and one refused further treatment. Additionally, one child died because of poor perioperative management of inflammation. None of the 13 surviving patients exhibited deterioration or distant metastasis during follow-up.

As IMT could potentially become malignant, CT and magnetic resonance imaging (MRI) examinations are recommended at 3 months, 6 months, and 1 year after resection. Subsequently, annual color Doppler ultrasound and CT or MRI examinations are recommended to detect recurrence and metastasis.

#### **Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

#### **Conflicts of interest**

None.

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