# Aggressive soft tissue tumours of infancy

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**BA Masri, QR Son-Hing, R Beauchamp. Aggressive soft tissue tumours of infancy. Can J Plast Surg 1994;2(2):81-84.**
Three cases of aggressive congenital soft tissue musculoskeletal neoplasms are presented. Two of these cases are congenital alveolar rhabdomyosarcoma of the foot. The third case is an infantile hemangiopericytoma of the thigh. These are rare conditions that should always be kept in mind when faced with a young infant with a mass in his or her limbs. Appropriate diagnosis and treatment are of paramount importance in these rare tumours. The management of these tumours is outlined and the pertinent literature reviewed.

**Key Words:** *Biopsy, Congenital sarconia, Infancy, Tumours*

**Tumeurs agressives des tissus mous chez le nourrisson**

**RÉSUMÉ :** Trois cas de néoplasie musculo-squeletique congénitale des tissus mous sont présentés. Deux de ces cas sont des rhabdomyosarcomes alvéolaires congénitaux du pied. Le troisième cas est un hémangiopéricytome infantile de la cuisse. Ce sont des maladies qui devraient toujours être gardées à l'esprit lorsqu'un nourrisson présente une masse au niveau des membres. La justesse du diagnostic et du traitement sont primordiaux dans les cas de ces tumeurs rares. On présente ici le traitement de ces tumeurs et un survol de la littérature à ce sujet.

Despite the fact that congenital fibrous tumours, particularly the fibromatoses, are second in frequency only to vascular tumours (1), little attention has been given to these tumours in the literature. Moreover, congenital malignancy of soft tissues has received equally little attention. Congenital sarcomas such as rhabdomyosarcoma, fibrosarcomas, malignant mesenchymomas, leiomyosarcomas and malignant endothelial hemangioendotheliomas have been reported in the literature (1-3). Unfortunately, there has been little emphasis on congenital sarcomas in the recent peer reviewed literature, and even in standard textbooks of plastic surgery, orthopedics and musculoskeletal oncology.

The purpose of this article is to report on congenital sarcomas in two children, and on a locally aggressive benign congenital neoplasm in a third patient, and to outline our approach to these problems.

## CASE PRESENTATIONS

Case 1: A 23-month-old boy presented because of an enlarging mass in the left heel. This was a previously healthy boy who was born to a gravida 4 para 3 healthy mother following a normal pregnancy and uncomplicated delivery. He first presented with a noticeable mass on the lateral side of the heel at seven weeks of age. A diagnosis of a congenital lymphangioma was made and no specific treatment was offered. The mass continued to enlarge and about four months before presentation it became painful, and the child began to limp. At the time of presentation, he was no longer able to walk because of pain, and the mass had enlarged to the point that shoes were difficult to fit. There was no history of weight loss, anorexia or fever.

Physical examination revealed an irritable child in obvious discomfort. There was a large mass extending from the lateral side of the heel into the calcaneus and the medial side of the heel (Figure 1). Range of motion of the ankle and subtalar joints were restricted. The mass was rubbery in consistency and was fixed. It did not transilluminate and did not transmit a bruit. Examination of the groin revealed marked lymphadenopathy. Plain radiographs did not show any significant bony pathology. A magnetic resonance imaging (MRI) scan was obtained and confirmed the invasive nature of the tumour (Figure 2). A metastatic workup revealed metastases to the left inguinal lymph nodes, to the right kidney and to bone marrow. The tumour and metastases were biopsied and revealed a high grade alveolar rhabdomyosarcoma. A palliative below the knee amputation was performed The amputation stump healed without incident, and the patient was then treated with postoperative chemo-

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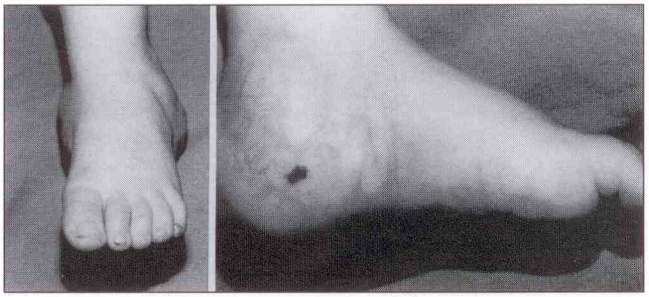

**Figure 1)** *Large tumour occupying most of the heel region in case 1. The tumour has grown to the point that the skin on the medial side of the heel was ulcerated. Because of the extent of the lesion, limb salvage was not possible and a below the knee amputation was performed (Case 1)*

therapy. At last review, three months after surgery, he was doing well with the chemotherapy and was walking very well with a below the knee prosthesis.

**Case 2:** A four-week-old girl presented for evaluation of a mass in the plantar aspect of her forefoot. She was born at term following a normal pregnancy and forceps delivery to a primigravida 22-year-old mother. There were no medical or developmental problems until the time of presentation. The mass was noticed by her mother during bathing.

On examination, she had a hard 2.5 cm nodule on the plantar aspect of her midfoot overlying the bases of the metatarsals and extending up to the level of the navicular. Following a computerized tomographic (CT) scan (Figure 3) and a negative metastatic workup, an open biopsy was performed. The biopsy revealed a high grade alveolar rhabdomyosarcoma. A Syme's amputation was performed at seven weeks of age and the patient was fitted with a prosthesis several months later. Postoperative chemotherapy was used as adjuvant treatment. At last follow-up, one year after surgery, she was well with no detectable tumour. She was walking well with her prosthesis.

**Case 3:** A three-week-old girl presented with a 9x5x5 cm mass in her left thigh, which was first noted at birth. This mass was biopsied inconclusively at age 10 days, and for this reason the patient was transferred to our hospital. Following a complete oncological work-up, including an MRI scan (Figure 4), bone scan and a CT scan of the chest, an open biopsy was performed. The biopsy proved to be an infantile hemangiopericytoma. Because of the high risk of local recurrence of such tumours, a wide resection was performed at the age of four weeks. Examination of the resection specimen's margins revealed no tumour and chemotherapy was not used. Following surgery, she was fitted with a knee orthosis. At three months, she was doing well, with no signs of tumour recurrence, and with only a slight persistent flexion deformity of her knee. Her only disability was quadriceps weakness on that side.

## DISCUSSION

Despite the paucity of reports on congenital sarcomas in the recent literature (4), many reports on this topic have been published in the past (1,5-8). Although the musculoskeletal system is the most common site for these tumours, congenital sarcomas have been reported in other regions (2,5,9). Hartz and Guerrero Tablante (6) reported the case of a congenital sarcoma in the mastoid region in a newborn girl. Ambramowsky and Witt (9) reported the case of a newborn infant with a poorly differentiated laryngeal sarcoma. Lazarus et al (2) reported the case of a nine-day-old infant with a large intrapericardial undifferentiated sarcoma.

The relative frequency and distribution of congenital aggressive neoplasms are difficult to determine because of the

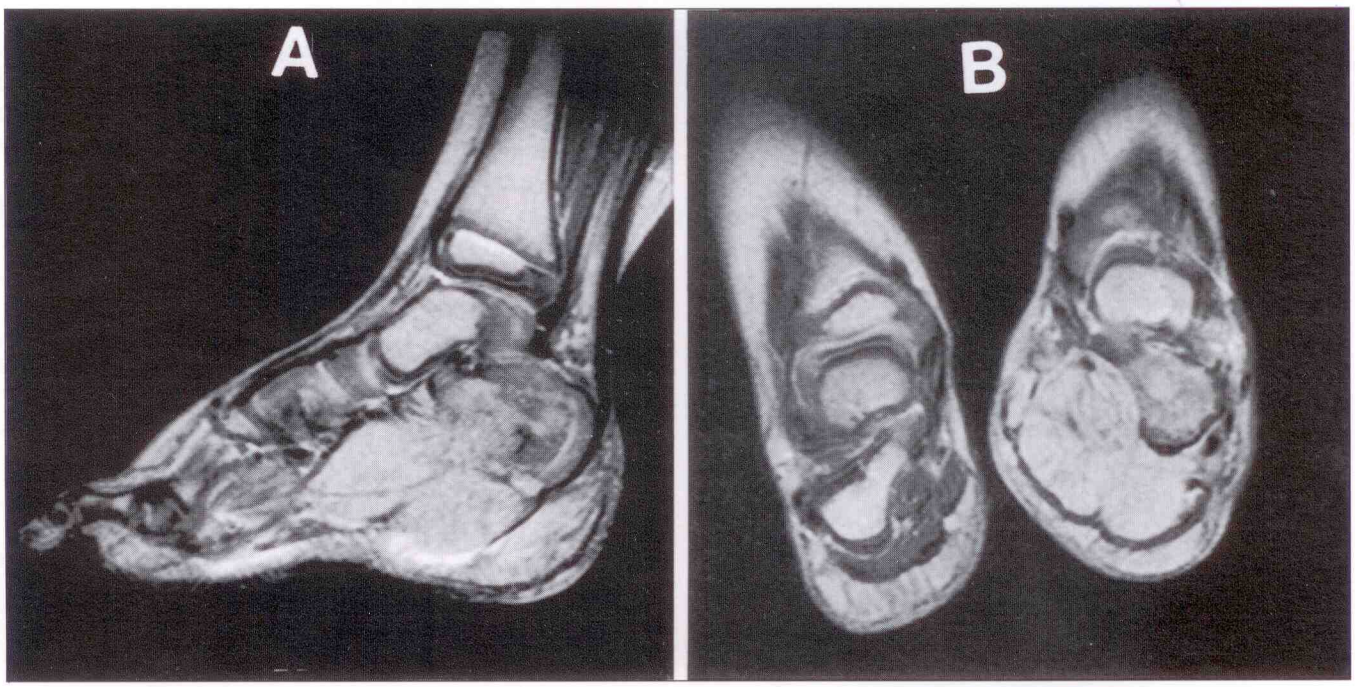


**Figure 2)** *MRI scan of the lesion shown in Figure 1. The coronal cuts* **(B)** *show the normal foot (left) compared with the abnormal foot (right). Note the massive size and degree of invasion into the calcaneus*

limited literature available. In a large review of congenital mesenchymal tumours, Kauffman and Stout (1) were able to accumulate 120 of these cases. The most common tumour in that series was fibromatosis. At least 50% of such cases affected the head and neck region. Only 10 out of 37 congenital fibromatoses affected the limbs. Although these are classified as nonmalignant neoplasms, their treatment is not unlike that of malignant tumours because of their aggressive nature and tendency towards local recurrence. Although a few of these tumours may regress spontaneously, most do not. Many of these tend to infiltrate widely, thus making a wide resection very difficult. Most local recurrences are due to inadequate excision.

In Kauffman and Stout's series (1) there were 15 congenital rhabdomyosarcomas, 11 malignant mesenchymomas, four fibrosarcomas and two malignant hemangioendotheliomas. Of the 15 congenital rhabdomyosarcomas (1), six involved the head and neck region, three involved the urogenital system, three involved the upper limb and perineum, one involved the lower extremity, and one involved the trunk. Six of these 15 infants died as a result of their tumour, eight survived with no recurrence and one was lost to follow-up. Of the 11 malignant mesenchymomas, six involved the limbs and five involved the trunk. Of these 11 patients, only one died as a result of his tumour. The authors felt that the presence of rhabdomyosarcoma, chondrosarcoma or angiosarcoma, in that order, was a poor prognosticator in malignant mesenchymomas. Of the four congenital fibrosarcomas, two were located in the foot and one was in the trapezium muscle. Both foot tumours were treated with an amputation, with no recurrence of the tumour and no distant metastasis. The trapezium fibrosarcoma was treated with piecemeal excision, and the result was death at six months of distant metastases. The two malignant hemangioendotheliomas were probably not true congenital malignancies. These were most likely malignant degeneration of congenital benign hemangioendotheliomas, a common congenital neoplasm.

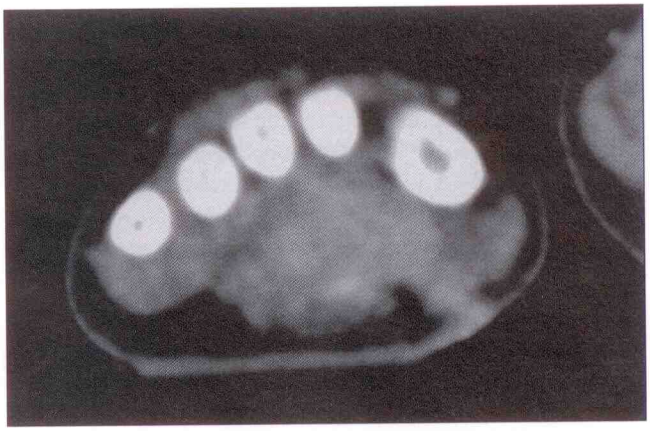

**Figure 3)** *CT scan of the foot rhabdomyosarcoma discussed in case 2. The tumour occupies most of the central compartment of the foot*

Two of the cases presented in the present report were congenital rhabdomyosarcomas occurring in the foot. This is consistent with Kauffman and Stout's series (1) in which rhabdomyosarcoma was the most common malignant congenital soft tissue tumour. In our first case, the congenital rhabdomyosarcoma was mistaken for a lymphangioma. This tumour did not have the characteristic appearance of a lymphangioma at the time of presentation to our clinic, neither did it behave in a manner suggestive of a lymphangioma. The steady growth of the tumour, as well as the presence of pain, should suggest that this is a more significant neoplasm. If

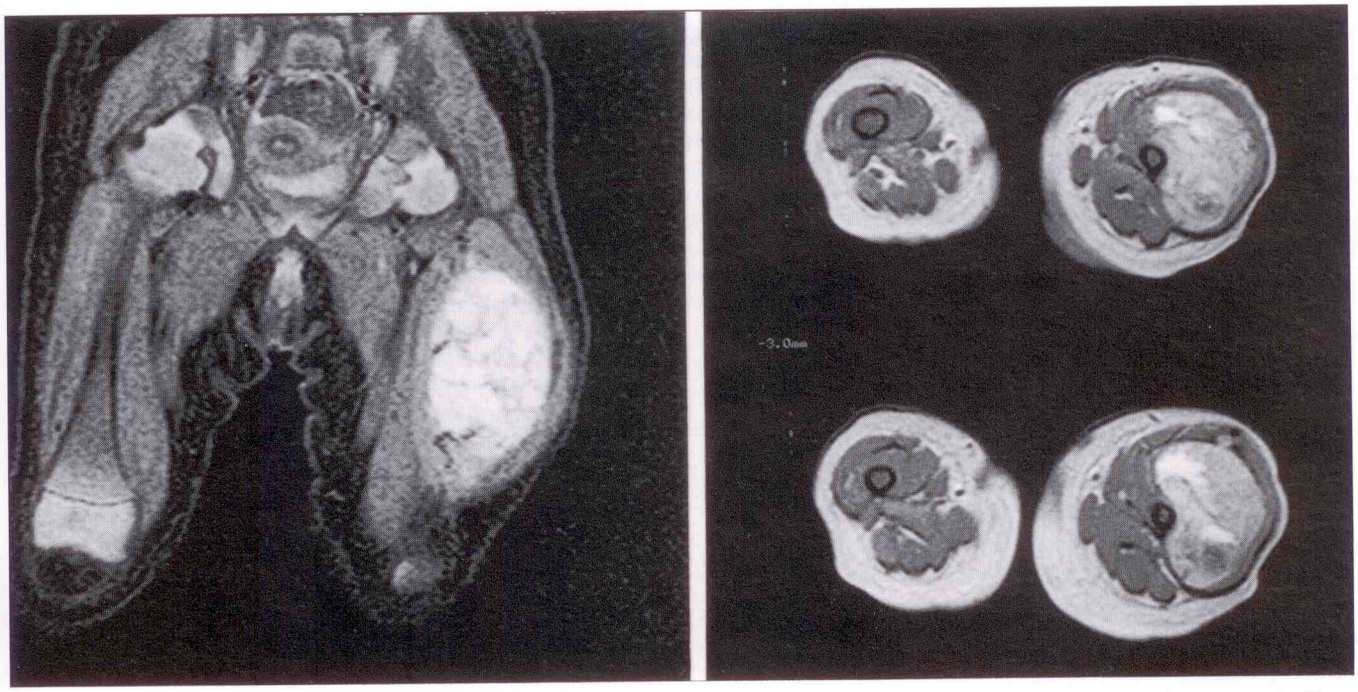

**Figure 4)** *MRI scan of the thigh hemangiopericytoma discussed in case 3. The tumour lies within, and occupies most of, the quadriceps muscle. There was no bony invasion*

there is any doubt about the diagnosis of a hemangioma, lymphangioma or any other mass in a child, an MRI study is mandatory for the complete investigation of the patient. If there is any doubt about the diagnosis, appropriate and prompt referral to a musculoskeletal oncological surgeon should be made. A biopsy in such a case should only be performed by an experienced musculoskeletal oncological surgeon, because inadequate biopsy technique may compromise the result and preclude the possibility of limb salvage surgery (10). The best treatment for congenital sarcomas continues to be a prompt wide resection, if possible. If the location precludes a wide resection, as was the case in our two patients, and in Kauffman and Stout's two patients with congenital fibrosarcoma of the foot (1), an amputation should be performed.

The third case presented in this report is not a true malignancy. The locally aggressive behaviour of infantile hemangiopericytomas deserves aggressive treatment in order to minimize the risk of local recurrence. Other locally aggressive congenital benign soft tissue tumours include the fibromatoses (11) and dermatofibrosarcoma protuberans (12).

We recommend that any soft tissue mass in a child that does not have the classic appearance of a capillary or cavernous hemangioma should be investigated. The investigation should be performed by an experienced musculoskeletal oncological surgeon, and should include an MRI scan and a thorough metastatic work-up. At our centre, this includes a CT scan of the chest, a technetium bone scan, and a thallium nuclear scan in addition to the standard investigations. Following completion of the metastatic work-up and staging investigations, a well planned biopsy should be performed. We prefer a needle biopsy if at all feasible. If this is not successful, we proceed with an open biopsy under the same anesthetic.

The basic tumour biopsy principles should be strictly adhered to. A tourniquet may be used; however, the limb should be exsanguinated with elevation only, and not with stripping. The incision or needle approach should be in line with the incision planned for a wide resection of the tumour, so that the biopsy tract may be completely excised at the time of tumour resection. Dissection should proceed directly towards the tumour, with minimal elevation of skin and subcutaneous tissue flaps. The tourniquet should be deflated prior to wound closure, and hemostasis should be meticulous. If a drain is needed, it should exit in very close proximity to the wound, and in line with the surgical incision. The specimen should be sent to the pathology laboratory fresh and not in formalin. Prior discussion of the case with the pathologist is very helpful. A rush section should always be obtained in order to ensure that an adequate specimen is obtained, and not simply tumour necrosis. The frozen section will also help the pathologist to determine whether the submitted specimen is adequate for all the special tests needed to make a definitive diagnosis, thus avoiding a second biopsy. After a definitive diagnosis is made, and if the tumour is malignant, the patient should be seen by a pediatric oncologist, for consideration of perioperative chemotherapy, if the tumour is sensitive to chemotherapy. The surgical treatment of choice is a wide resection, if feasible. If the location of the tumour precludes a wide resection, an amputation should be performed. There is no role for a piecemeal excision of the tumour.

In summary, the possibility of a mass noted within the first few weeks of life being a congenital malignancy, or a locally aggressive benign neoplasm, should always be kept in mind. Because of the rarity of these conditions, a prompt diagnosis requires a high index of suspicion, and any delay in diagnosis or treatment may compromise the patient's final outcome.

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