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CASE REPORTS

Eight-year follow-up after scapulectomy in a neonate with congenital Ewing sarcoma of the scapula

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With an incidence of 2 per 100,0000, Ewing sarcoma represents the second most common primary bone neoplasia after the osteosarcoma.[4] Only 2.6% of cases occur by the age of 3.[9] The classic peak lies between ages 15 and 30 years. Only 2 publications have reported the existence of congenital skeletal Ewing sarcomas.[5,6] To date, a treatment protocol with a curative course has not been described in the literature. Although histopathology seems to be inconsistent, genetic examination usually reveals a typical chromosomal translocation.[1] In this report, we present the diagnosis, treatment with neoadjuvant chemotherapy and operative resection, and clinical and radiologic follow-up after 5 and 8 years postoperatively.

Case report

Diagnosis and neoadjuvant chemotherapy

On February 12, 2008, a male neonate in good general condition was born by primary cesarean section in the 38th week of pregnancy (weight, 3770 g; length, 54 cm). The Apgar score at birth was 10/10. At clinical examination, the newborn presented a 4- × 3-cm large, painless, firm, and elastic supraclavicular mass. The suspected diagnosis primarily was a congenital fibromatosis.

A sample from an excision biopsy was obtained 2 days after magnetic resonance imaging (MRI). The primary histologic diagnosis was a fibrous myomatosis. Further histologic analysis revealed CD99 expression; therefore, an Ewing tumor was suspected. On February 25, 2008, the fourth reference pathology revealed a translocation-positive (t7-22) congenital Ewing tumor with gene fusion transcripts type EWS-ETV 1.

Immediately afterward, a freely adapted chemotherapy following the Cooperative Weichteilsarkom Studiengruppe (CWS) protocol was initiated. Tumor staging did indicate distant metastases.

From MRI imaging on March 7, 2008, an initial tumor volume of 32.8 ml was estimated. A control MRI on May 19, 2008 revealed a remaining volume of 4.3 mL (reduction of 80%), and on June 12, 2008 the mass had decreased to a volume of 1.6 mL (reduction of 95%). A regression grade 5 according to Salzer-Kuntschik was therefore achieved (Fig. 1).[13] The operative resection of the Ewing tumor by scapulectomy was performed (Fig. 2) on June 18, 2008.

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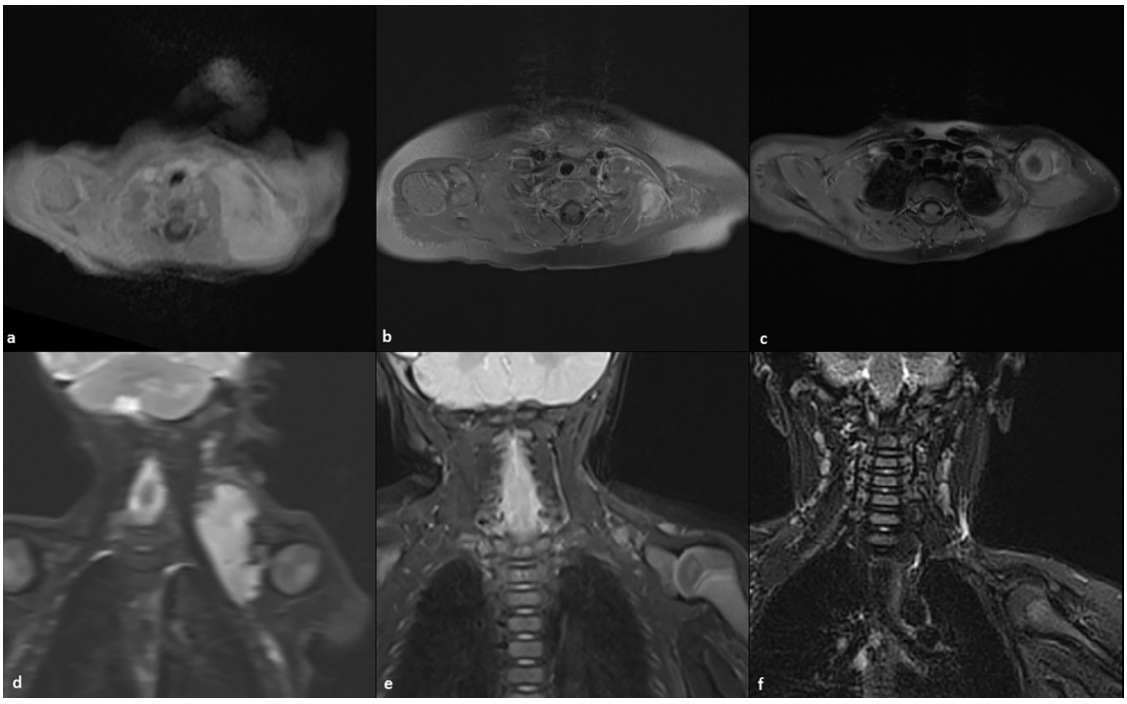

Figure 1 Magnetic resonance imaging (MRI) documenting tumor volume reduction throughout neoadjuvant chemotherapy. Axial, T1-weighted, contrast-enhanced images on (**a**) March 7, 2008, (**b**) June 12, 2008, and (**c**) June 20, 2011, as well as coronal, T2-weighted images on (**d**) March 7, 2008, (**e**) June 12, 2008, and (**f**) August 28, 2009.

Surgical technique

The operation took place with the neonate in a lateral position. The incision was performed taking into account the scar of the sample biopsy excision, starting semicircularly, dorsally along the medial margin of the scapula and reaching ventrally to the coracoid process. In this respect, it is important that sample excision and tumor resection are performed by the same surgeon. The resection line was oriented at the initial tumor size before neoadjuvant chemotherapy.

After preparation of the subcutaneous tissue, first, the deltoid muscle and the trapezius muscle were detached of acromion and spina scapulae. In the next step, the scapula was mobilized by separating the medial margin of the rhomboids at the scapular medial margin, beginning at the inferior angle. The scapula could then be detached medially, allowing the exposition of the thoracic wall.

The humerus was approached from the dorsal side. In this regard, the scapular inferior angle was rotated medially and upward to be able to detach the teres major muscle from the humerus. At the greater tuberosity, the supraspinatus, infraspinatus, and the teres minor muscles were detached close to the tendon insertion, and the glenohumeral joint was inspected. Between the teres minor and teres major (ie, the dorsal axillary gap), the axillary nerve was carefully dissected and spared.

On the ventral side, first the deltoid muscle was resected from the acromion; afterward, starting in the rotator interval, tenodesis of the long biceps tendon was performed, and the subscapularis muscle was detached at the lesser tuberosity. Anteromedially the levator scapulae muscle was dissected and cut off. The short head of the biceps muscle, coracobrachialis, and pectoralis minor muscles were detached from the coracoid process, while sparing the musculocutaneous nerve, and were sheathed to prepare reinsertion. The coracoacromial and coracoclavicular ligaments were also cut off from the coracoid process to release the scapula. Finally, the acromioclavicular joint was separated; thus, the scapula could be removed in total and was prepared for histologic examination.

For reconstruction, the humeral head was positioned at the thoracic wall under the clavicle, which consequently served as the new bony arch of the shoulder. The trapezius and deltoid muscles were circularly looped around the humeral head from posteromedial to ventral and were attached transosseously to the clavicle. Afterward the conjoined tendons and the pectoralis minor muscle were also reinserted to the clavicle.

Outcome and follow-up

Histologic analysis showed tumor-free resection margins (R0 resection) with a minimal distance of 0.4 cm and 9 tumor-free lymph nodes. Offshoots of the above-described Ewing tumor at the costal surface of the resected scapula could be observed.