Image Findings and Pre-operative Embolization of Pediatric Ewing’s Sarcoma of Skull: a case report

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Ewing’s sarcoma of the skull is a rare and aggressive primary bone tumor occurs in young-aged patients. The current main therapeutic strategy is surgical tumor removal followed by adjuvant chemoradiotherapy. We, hereby, present a pediatric case of Ewing’s sarcoma arising from right temporal bone. Pre-operative embolization of the hypervascular tumor followed by immediate surgical resection of the lesion was performed. The surgery resulted in a near-total tumor removal and a small residual tumor at the adjacent meninge. Early recurrence of the small residual tumor developed shortly after the surgery. This short time interval indicated the importance and challenge of complete resection for a malignant tumor of skull.

Ewing’s sarcoma is an uncommon primary bone tumor of childhood and adolescence, which account for around 10% of malignant bone neoplasm [1]. The most common location is metaphysis of long bones, pelvis and ribs. Ewing’s sarcoma of the skull is rare. The classical image findings of Ewing’s sarcoma of skull include large extra-axial tumor with or without bony destruction [2]. Ewing’s sarcomas of skull may present periosteal reaction and usually enhance vividly, suggesting hypervascularity [2]. Currently, the recommended treatments include radical tumor excision and chemoradiotherapy [3]. However, tumor hypervascularity makes surgery risky and massive bleeding during surgical resection unavoidable.

Pre-operative embolization is frequently used to facilitate surgery of hypervascular head and neck tumors [4, 5]. Hereby, we report a case of primary pediatric Ewing’s sarcoma of skull treated with combined preoperative embolization and immediate surgical resection.

CASE REPORT

A 5-year-old girl presented dizziness and vomiting for 3 days. Her general physical examination revealed a mass lesion at the right temporal region. Her pre-contrast brain computed tomography (CT) showed a large hypo-attenuated mass (63 x 48 x 54 mm in three dimensions) with marked “sunburst” periosteal reaction at the right sphenoid ridge and temporal bone. Strong and heterogeneous enhancement of the mass was observed after intravenous contrast agent administration (Fig 1a). On magnetic resonance imaging (MRI), a well-demarcated expansile bony tumor arising from right temporal skull was found. The tumor was hypointense on T1-weighted MR images (T1WI) and heterogeneous hyperintense on T2-weighted MR.
Images (T2WI) (Fig. 1b). Remarkable peripheral contrast enhancement was found after intravenous gadolinium-DTPA injection. A focal protruding enhancing nodule with suspicious invasion to the underneath meninges was noted (Fig. 1c). Cerebral angiography revealed the tumor hypervascularity with main feeding arteries from right middle meningeal and right ophthalmic arteries (Fig. 2a). High grade tumors, e.g. primitive neuroectodermal tumor (PNET), were impressed pre-operatively based on the image findings.

Pre-operative embolization for tumor devascularization and decrease surgical risk was attempted under general anesthesia. During the procedure, we navigated a microcatheter to the branches of right middle meningeal artery, and slowly injected 100-300µm Embosphere microspheres (BioSphere Medical, Rockland, MA) particles. Complete

**Figure 1.** a. A 5-year-old girl presented nausea and vomiting. Axial CT scan of brain, bone window, showed an expansile bony mass in right temporal region with marked “sunburst” periosteal reaction. b. MR image of brain, T2WI, showed a slightly hyperintense extra-axial mass. c. Post-contrast T1W MR image showed peripheral enhancement and focal protruding meningeal invasion (arrowhead).

**Figure 2.** a. Right carotid angiogram showed the tumor stain (arrowheads) fed by right middle meningeal artery. b. Control angiogram after embolization showed complete obliteration of tumor stain.
oblitration of tumor stain was achieved (Fig 2b). The tumor was operated immediately after embo-

Pathological examination of the excised tumor showed diffuse infiltration of small round cells with large nuclei and scanty cytoplasm. Some necrotic areas and hemorrhage were also noted. Immunohistochemical studies were positive for CD99, and negative for CD20 and LCA. The final diagnosis of Ewing’s sarcoma of skull was made accordingly.

Follow up MRI at 3 weeks after the operation showed an enhancing meningeal nodule adjacent to the operation site at right temporal region (Fig. 3). The location corresponded to the protruding enhancing nodule appeared initially on pre-operative MRI. Early tumor recurrence was impressed. The patient was then referred to oncology department for further chemotherapy. The patient was in stable condition in a 4-month follow-up.

**DISCUSSION**

Ewing’s sarcoma is the second (after osteo-
sarcoma) most common primary pediatric bone malignancy, accounting for about 10% of all malignant bone tumors [1]. It is more common in whites and rare in blacks and Asians. The tumor has predilection for metadiaphysis of long tubular bones although any bone of the body can be involved. An Ewing’s sarcoma occurred in skull is rare and accounts for only between 1 and 6% [6]. Frontal and parietal convexities are the most common locations of skull Ewing’s sarcomas. Temporal bone, ethmoid bone, orbit and sphenoid bones are the other less common sites [6, 7]. There is also no gender preference for Ewing’s sarcoma of skull. The median age of presentation of Ewing’s sarcoma is 13 years and about 75% of the patients are in their second decade of life [1].

Clinically, the most common symptoms of Ewing’s sarcoma include pain and swelling. The pain is usually intermittent but may increase in severity with time. Fever, anemia, increased erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), sometimes, also occur. Unusual presentations due to facial palsy or increased intracranial pressure caused by tumor have also been reported [8, 9]. Unlike Ewing’s sarcoma in tubular bones, pathologic fracture rarely occurs in primary skull lesions.

The image findings of skull Ewing’s sarcoma were reported before [10, 11]. It is usually hypo-
attenuated on CT. Typically, skull Ewing’s sarcomas are low-signal on T1WI and iso- to high-signal on T2WI extra-axial mass lesions. Necrosis or hemor-
rhage may occur. With the tumor origin from intra-
medullary cavity, bony expansion and destruction are common in Ewing’s sarcoma [8, 11]. Lamellated periosteal reaction known as “onion-peel”, typically to Ewing’s sarcoma of tubular bones, are rarely seen in skull Ewing’s sarcoma [11]. However, in our case, prominent periosteal reaction resembles “sunburst” or “hair-on-end” periosteal reactions, common in osteogenic sarcoma and less common in Ewing’s sarcoma in tubular bones, are present. This is the most striking image finding on plain radiography and CT. Similar finding was reported only once, to our knowledge [7]. The tumor itself appeared relatively hypo-attenuated on plain CT and heterogeneously enhanced after intravenous injec-
tion of iodinated contrast medium. On MR images, the current tumor appeared as an extra-axial mass arising from right temporal bone. The tumor was slightly hyperintense on T2WI and hypointense on
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TIWI. Enhancement after intravenous gadolinium-DTPA injection was noted at the periphery of tumor. Initially, the tumor was considered extra-axially located without invasion to the underneath brain parenchyma. Retrospectively, it is believed that the focal protruding nodule at the temporal cortical region may imply meningeal invasion. The local invasiveness was proved, afterwards, by operation. Although the tumor was considered “totally” extirpated initially, this subtle meningeal invasion became the origin of early tumor recurrence and seen on MRI 3 weeks after surgery.

Meningioma is one of the common intra-cranial extra-axial tumors that may benefit from pre-operative embolization [4, 12-13]. Based on image findings, initially, we considered the surgical procedure a high risk one with potentially massive blood loss in such a young patient. Conceptually, decrease intraoperative blood loss and surgical difficulty with shorter operation time are expected after preoperative embolization of a hypervascular head and neck tumor [4, 13]. However, the far majority of preoperative embolization was applied to meningiomas [5]. Technically, no much difference between the preoperative embolization of meningiomas and other head and neck tumors is presumed. We, therefore, used Embosphere, a small-sized hydrophilic, nonresorbable, collagen-coated, acrylic microsphere (100-300µm), for embolization and expected to achieve most distal arterial and maximal perfusion blockade. The goal seemed achieved in the current case. The optimal time interval between surgery and preoperative embolization is still disputable and may differ according to the embolization agents used. Kai et al. used cellulose porous beads as the embolization particles and reported the optimal time interval of 7-9 days [14]. Chun et al. used PVA particles and recommended surgical resection of meningiomas should be delayed for more than 24 hours after embolization [15]. In clinical practice, the time intervals between embolization and surgery were ranged from less than 1 day to several days or weeks. We chose surgery immediately after embolization for the current case mainly with the concerns of potential risk relevant to another session of general anesthesia.

CONCLUSION

In this report we described the image findings and pre-operative embolization of a child with Ewing’s sarcoma in skull. And because of the hypervascularity, we believe pre-operative embolization is a practical way to minimize potential massive blood loss. Complete resection is critical to the outcome and close post-operative follow-up is very important to detect early tumor recurrence.

REFERENCES

原發於小兒顱骨之尤恩氏肉瘤之影像學特徵與手術前栓塞療法：病例報告

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原發於顱骨的尤恩氏肉瘤為一種好發於年輕人的罕見惡性骨腫瘤。目前的治療方向主要是以根除性手術搭配放射線治療及化學治療為主。我們在此報告一例原發於小兒病人右側顱骨的尤恩氏肉瘤。栓塞治療之後我們立即搭配施行根除性手術。手術結果腫瘤近乎完全去除，然而在貼近腦膜處仍然有少量的殘存腫瘤。術後不久腫瘤就有早期復發的情況。這種短期內復發的情況表示對顱骨惡性腫瘤而言，徹底的根除性手術是有其重要性的。