Congenital Angiosarcoma, a Case Report and Review of Literature

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Abstract

Introduction: Angiosarcoma, originating from vessels, constitutes about 0.2% to 0.3% of all pediatric soft tissue sarcomas. Prognosis of angiosarcoma is poor and depends on patient’s age, tumor location, size, histological grade and extent of tumor progression.

Case Presentation: We report a rare case of a congenital angiosarcoma of scalp with dural and skull bone invasion in a one-month-old boy. His mother noticed the mass 2 days after birth as a very small insignificant nodule, but it grew rapidly afterward.

Conclusions: The treatment consisted of only a wide surgical resection. After 15 months there was no sign of local recurrence or metastasis was noticeable and the tumor showed favorable outcome. This case indicates the possibility of a better clinical behavior in congenital angiosarcoma.

Keywords: Angiosarcoma, Congenital Tumor, Scalp

1. Introduction

Soft tissue sarcomas are mesodermally derived malignant tumors from muscle, fat, cartilage, bone, nerve, fibroblast and vessels, accounting for about 8% - 10% of all cancers of children or young adults (1). Angiosarcoma, originating from vessels, constitutes about 0.2% to 0.3% of all pediatric soft tissue sarcomas (2, 3). It can occur in any site in the body, however the majority of adult cases have been reported in skin and superficial soft tissue of head and neck region (4) but in children most reported cases originated from mediastinum, pericardium and heart (5). There are also rare case reports of congenital and intrauterine angiosarcoma (6).

Angiosarcomas have been reported as aggressive tumors with poor prognosis (7), however the prognosis depends on factors such as tumor size, grade, and patients’ age. In this article we report a rare occurrence of a congenital angiosarcoma of scalp with dural and skull bone invasion and a favorable outcome.

2. Case Presentation

A one-month-old boy born by caesarean section presented with large scalp mass (3 × 3 cm). His mother noticed the mass two days after birth as a very small insignificant nodule. The nodule grew rapidly and he was referred to our center at one month of age. Patient’s birth weight was 3100 grams and he was a well baby without any other problem.

Physical examination was unremarkable except the mentioned scalp mass measuring about 3 × 3 cm in the left parietal area.

Parents had a history of infertility for 10 years and both of them had received gonadotropin-releasing hormone agonist (GnRH). The father had multiple sclerosis for 15 years and was treated with interferon.

Magnetic Resonance Imaging (MRI) and CT scan of the skull displayed a well demarcated 28 × 28 mm mass in left parietal bone with intracranial extension and pressure effect on adjacent brain tissue (Figure 1A).

Laboratory data were unremarkable except for hemoglobin of 10 g/dL. Renal as well as liver function tests were normal.

No evidence of any vascular malformation or tumor was detected in other sites of the body.

The bulging red mass was excised by neurosurgeon. At the time of first surgery the tumor showed obvious dural invasion and underlying dural perforation (Figure 1B).

Microscopic examination of multiple fragments of bone and soft tissues showed a highly vascular tumor with hyperchromatic atypical nuclei and many mitotic figures (about 6 per 10 high power fields (HPF)) with focal microscopic necrosis. Bony tissue labeled as temporal bone was...
Figure 1. A, CT scan image showed a soft tissue mass with obvious bone and dural involvement; B, image of bulging red subcutaneous mass in the scalp before resection; C and D, highly vascular tumor with anastomosing vessels in C lined by atypical cells with hyperchromasia and mitosis in D (Hematoxylin and Eosin stain); E and F, result of IHC study: membranous staining of tumor cells by CD31 marker (E) and high proliferative index by ki67 nuclear staining of tumor cells (F).

also involved by tumor. The vascular spaces were interconnected and abnormal (Figure 1C and D).

Immunohistochemistry (IHC) showed positive CD34 and CD31 and high proliferative rate (ki-67 of 60% - 80%) (Figure 1E and F).

The patient underwent another surgery and a segment of dura which has been invaded by the tumor was resected again. The dural tissue showed macroscopic and microscopic invasion by tumor with the same morphology.

According to the large size, high mitotic figure and
differentiated tumor (G1) and was in T2a stage in opposite
with regard to its rarity.

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phedema associated AS (13).

Deep Soft tissue AS, visceral organ AS, radiation and lym-
on the clinical presentation and tumor site: cutaneous AS,
ies of these 5 congenital cases are listed in Table 1.

Angiosarcomas (AS) are classified in five groups based
on the clinical presentation and tumor site: cutaneous AS,
deep Soft tissue AS, visceral organ AS, radiation and lym-
phedema associated AS (13).

Rapid growth and invasion to adjacent tissue, pre-
cludes exact determination of the tumor origin. However,
its most common location in adults is skin and superficial
soft tissue of head and neck (14). In children case reports
are very rare, however, here too the favorite location seems
to be head, neck and mediastinum (8).

Cutaneous AS presents as purple nodule with or with-
out ulceration (15).

In our patient, the tumor was located in the head and
neck (scalp area). It was first detected by the mother and
appeared to have originated from the soft tissue with inva-
sion to bone and dura. Benign vascular lesions are more
common in children and important malignant tumors
such as rhabdomyosarcoma should be ruled out. Rhab-
domyosarcoma is a small round cell tumor and is nega-
tive for vascular markers like CD31 and CD34. Hemangoen-
dotheiloma has not the branching anastomosing vessels
that is typical of angiosarcoma. In conclusion, clinical
behavior of the tumor, imaging of the patient with du-
ral extension and histologic pictures of the tumor with
branched intercalating vessels lined by highly atypical
cells and presence of many mitosis and the result of IHC
study with high Ki67 and positivity of tumor cells for vas-
cular markers all helped us to diagnose angiosarcoma even
with regard to its rarity.

According to AJCC, TNM system, our patient had a well
differentiated tumor (G1) and was in T2a stage in opposite
to most of the angiosarcomas that were in an advanced
stage at presentation.

Because of the rarity of this tumor in children, infor-
mation about the long term effect of treatment modalities
is sparse. Currently, the most commonly used treatment
is wide surgical excision. If the complete excision is not
unachievable, radiotherapy and sometimes chemother-
apy can be started. Radiotherapy is more commonly used
when the margins are involved to prevent local recurrence.
The effect of chemotherapy is still unclear (14, 16).

Our patient underwent complete surgical excision in
two steps without any incident and his parents did not
give their agreement for chemotherapy or radiotherapy.
However, surprisingly, he is alive so far (15 months after
surgery). He is symptom-free but parents did not give their
consent for imaging study in follow up.

Overall prognosis of angiosarcoma is poor and de-
ends on patient’s age, tumor location, size, histological
grade and extent of tumor progression (7). The reported
outcome in children is also poor but our case and two
other cases of congenital cerebral angiosarcoma reported
by Suzuki et al. (9) and Kirk et al. (10) had good prognosis
even without any other treatment except for wide surgical
excision.

In the study performed by Ferrari et al on 18 patients
with malignant vascular tumors, 4 patients had no recur-
rence after 5 years. Size of tumor in 3 of them was less than
5 cm, so complete surgical excision was feasible and no fur-
ther treatment was instituted in the patients (17).

Our case and above mentioned congenital cases of an-
giosarcoma with good prognosis were also less than 5 cm
in size and were treated solely with wide surgical resection.
Thus, the small size of tumor and feasibility of complete
surgical resection can be considered as good predictors of
favorable outcome in these patients.

This case depicts a favorable clinical behavior of con-
genital angiosarcoma, however the number of cases in the
literature are still very few to make a definite suggestion.

Footnotes

Authors’ Contribution: Study concept and design: MS
and BG. Acquisition of data: MS, BG, BS, and MM. Draft-
ing of the manuscript: MS and BS. Critical revision of the
manuscript for important intellectual content: BG. Patient
final diagnosis: BG and MS. Patient treatment: MM

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lose.

Informed Consent: Informed consent was obtained from
the patients’ parents.
Table 1. Clinicopathological Characteristics of Congenital Angiosarcomas Reported so Far

| Year      | Age          | Sex    | Clinical Presentation          | Tumor Location and Size | Size, cm | Treatment          | Prognosis                          | Our Patient                          |
|-----------|--------------|--------|--------------------------------|-------------------------|----------|-------------------|-----------------------------------|--------------------------------------|
| Mena et al. [11] | 1999         | NR     | Enlarging head, irritability, poor feedings, vomiting and right proptosis | Intracranial            | NR       | Surgical excision | Died after 26 months with no sign of recurrence | Died at 34 days of age                 |
| Kirk et al. [10] | 1992         | Female | Enlarged head and a 1-week history of projectile vomiting | Intracranial            | 2.5      | Wide surgical excision | Alive after 11 months with no sign of recurrence | Alive after 15 months with no sign of recurrence |
| Suzuki et al. [9] | 2000         | Male   | Parotid swelling with an overlying wound since birth | Face from skull base to upper mediastinum | NR       | Chemotherapy (vincristine, cyclophosphamide, prednisone, paclitaxel) | Died at 7 months of age                | Died at 33 months after surgery         |
| Good et al. [6]  | 2008         | Female | Face deformity, ear discharge, respiratory distress, lung metastasis | Arm soft tissue | 6       | Partial surgical excision | Died at 34 months of age            | Died at 34 months after surgery         |
| Qureshi et al. [12] | 2012         | Male   | Rapidly growing swelling with an overlying wound since birth | Scalp soft tissue | 3       | Wide surgical excision | Alive after 15 months with no sign of recurrence | Died at 33 months after surgery         |

Abbreviation: NR, not reported.

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