Extraskeletal Ewing Sarcoma involving abdominal wall in a neonate

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ARTICLE INFO

Article history:
Received 21 November 2013
Accepted 26 November 2013
Available online 22 December 2013

Key words:
Extraskeletal Ewing Sarcoma
Abdominal wall
Congenital

ABSTRACT

Ewing Sarcoma is the second most common primary malignant bone tumor in children and young adults following osteosarcoma, but exceptionally it can arise from extraskeletal sites also. Extraskeletal Ewing Sarcoma is of neuroectodermal origin and usually involves extremities, retroperitoneum and para-vertebral regions. In literature, we have found only two case reports of Extraskeletal Ewing Sarcoma involving abdominal wall in adults. We present a neonate who was born with a mass at left anterolateral abdominal wall which turned out to be Ewing Sarcoma. To the best of our knowledge, this is the first case of Extraskeletal Ewing’s Sarcoma in a neonate involving abdominal wall.

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Ewing sarcoma is a bone neoplasm that usually occurs in children and young adults but rarely it can arise from soft tissue, labeled as Extraskeletal Ewing Sarcoma. We present a neonate who was born with a mass at left anterolateral abdominal wall which turned out to be Ewing Sarcoma and patient is now in 12th month follow up at Paediatric Oncology Department.

1. Case report

A 7 days old, full term male neonate, born through caesarian section at a District hospital and was referred to us with complaint of swelling left anterolateral abdomen since birth. There was no history of fever, GIT or urinary complaints and respiratory distress. Patient had birth weight of 2.5 kg and there was no history of any drug intake by mother during gestation. On examination patient was pale with normal vital signs. Examination of swelling showed a round multi-lobed swelling measuring 15 × 10 cm present on left anterolateral aspect of abdomen with overlying veins and no obvious signs of inflammation; however there were multiple areas of redness over the swelling. Swelling was nontender with normal overlying temperature and not transilluminant. It was fixed to the underlying tissues extraperitoneal, irreducible and had adherent overlying skin. His laboratory reports showed anemia (Hb: 7.2 mg/dl). Ultrasound of the swelling showed a mass in left flank with high velocity blood flow with suspicion of lymph-hemangioma. His MRI showed that bulk of the mass was within the subcutaneous tissue with stretching of the overlying skin and no infiltration into the underlying and deep structures of the abdomen noted. The mass was isointense on T1W1 with few hyperintense foci, mixed signal intensity on T2W1 and STIR. Our radiologist had a suspicion of Teratoma, so his AFP levels were advised but that was also within normal range. His excision biopsy was planned and patient was optimized after blood transfusion. Peroperatively mass was extending to abdominal wall muscles only with no intraperitoneal extension. Mass was excised completely and muscles were repaired. Skin defect was closed by skin grafting. His postoperative recovery remained uneventful. His biopsy report showed it to be round blue cell tumor which was further labeled as Ewing Sarcoma after immunohistochemicals staining (CD 99). His metastatic workup including bone scan was obtained which turned out to be normal. His CT chest showed few indeterminate subpleural nodules measuring 2–3 cm in both lungs. Patient was referred to Paediatric Oncology Department for further management. Patient was lost in follow up for 6 months and then he presented to Paediatric Oncologist for management. Oncologist decided to observe the patient and not to start chemotherapy. Patient is in our strict follow up and is now 1 year old; and is doing well to date.

2. Discussion

Extraskeletal Ewing Sarcoma (EES)/Primitive Neuroectodermal Tumor (PNET) are rare lesions which are of soft tissue origin. They usually originate from extremities, retroperitoneum, chest wall and...
paravertebral regions. Less common sites include pelvis, hip, bladder and rarely abdominal wall. They usually affect young adolescents and adults between age of 10 and 30 years with slight male predominance. They most commonly present as mass in soft tissues but it varies and depends upon site of origin [1].

EES is of neuroectodermal origin and one of the round blue cell tumors. Classically it is composed of small round cells, arranged in sheets and blue in color. The cells have high N/C ratio and vacuolated cytoplasm because of presence of glycogen; nuclei are round with “salt & pepper” chromatin [2]. Further definitive distinction into Ewing sarcoma/PNET requires usage of specific immunohistochemical markers like CD99 (MIC-2). This family of tumors shares common cytogenetic and molecular changes that involve translocation of the ES gene on chromosome 22 (22q12) onto several other genes such as FLI-1 on chromosome 11 (11q24) in 90% of cases and rearrangement on chromosome 21 (21q22). In addition, it appears that patients carrying the transcript EWS/FLI-1 have slower tumor growth than other transcripts and better response to chemotherapy [3]. Radiological diagnosis might require Ultrasound, CT and MRI with nonspecific findings [4].

The treatment of EES involves both chemotherapy and radiation therapy along with surgical resection. The consistent use of chemotherapy regimen in treatment of localized disease has significantly increased 5 and 10-year survival rates [5]. The prognosis of EES is considered better than Ewing Sarcoma of bone [6]. Various drugs had corresponding survival rates of 47% and 31% [8]. Of 66% and 61% at 5 and 10 years compared to older patients with Ewing Sarcoma [7]. Age at the time of diagnosis is considered a good prognostic factor [2].

EURO.E.W.I.N.G have been using 6 courses of Vincristine, Ifosfamide, Etoposide (VIDE) as induction chemotherapy [6]. Radiological diagnosis might require Ultrasound, CT and MRI with nonspecific findings [4].

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The common feature among those of survivals in their series was control of disease, it was a real tough question whether to go for chemotherapy or not. In our case, oncology board decided to wait for follow up and not to start treatment because of the hazards of chemotherapy and radiotherapy at this age (Figs. 1–3).

Two case reports of EES involving abdominal wall have been found which were of patients of age of 35 and 65 years, but no case of EES in neonates involving abdominal wall has yet been reported [2,10].

3. Conclusion

Congenital Extraskeletal Ewing sarcoma is although rare but an important entity. Complete surgical excision with safe surgical margins should be considered a primary treatment but decision for chemotherapy varies from case to case.

References