

A Soft Tissue Aneurysmal Bone Cyst in Thoracic Cavity

Torasik Kavite Yerleşimli Yumuşak Doku Aneurizmal Kemik Kisti

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Abstract

Objective primary aneurysmal bone cyst is a benign, locally aggressive and expansive tumor that typically occurs in long bones or vertebral bodies of children and young adults. Aneurysmal bone cysts originating from the soft tissue are extremely rare, and there has been only one case of soft tissue aneurysmal bone cyst located in the thoracic region. Almost all cases of intrathoracic aneurysmal bone cyst are associated with ribs. We describe a case of primary aneurysmal bone cyst of the soft tissue in the thoracic cavity of a 15-year-old boy. Although initially the tumor was considered cyst hydatid on radiological imaging, microscopic features of the tumor were identical to those of classical aneurysmal bone cyst. Intrathoracic aneurysmal bone cyst, an associated with ribs is extremely rare, due to aneurysmal bone cyst originating from the soft tissue of thoracic cavity, however, radiological and pathological data along with the evaluation can be diagnosed.

Key words: Aneurysmal, bone, cyst.

Özet

Primer aneurizmal kemik kistleri; çocuk ve genç erişkin dönemde, tipik olarak uzun kemikler ve vertebra gövdesinde görülen, benign, lokal agresif ve genişleyebilen tümörlerdir. Yumuşak dokudan kaynaklanan aneurizmal kemik kistleri oldukça nadirdir ve literatürde toraksik kavite içinde yumuşak dokudan kaynaklanan kemik kisti yalnızca bir olguda bildirilmiştir. Çoğunlukla torasik kavite aneurizmal kemik kistleri kosta ile ilişkilidir. Bu makalede, 15 yaşında erkek çocukta saptanan ve toraks boşluğunda gelişen, yumuşak dokunun primer aneurizmal kemik kisti sunulmuştur. Olgu radyolojik olarak öncelikle kist hidatik olarak düşünülmüş, mikroskopik bulguları ile klasik aneurizmal kemik kisti ile uyumlu bulunmuştur. İntratorasik yerleşimli, kemik ile ilişkisi olmayan aneurizmal kemik kisti olgusu son derece nadirdir, bu olgunun yumuşak doku kaynaklı olduğu ancak radyolojik ve patolojik verilerinin birlikte değerlendirilmesi ile belirlenebilir.

Anahtar Sözcükler: Aneurizmal, kemik, kisti.

Primary aneurysmal bone cysts (ABCs) are benign, locally aggressive, and expansive tumors that typically occur in the long bones or vertebral bodies of children and young adults (1,2). They occur exclusively within the bones. Primary ABCs developing in the soft tissue (STABCs) is extremely rare; however, in recent years, a few cases of STABCs located in the upper extremities, thighs, groins, shoulders and abdominal wall have been also reported

(3-6). The only case of a STABC located in the thoracic cavity and not associated with bones was a fourteen-year-old child (6). Few cases of ABCs located in this region have been reported, and all of them are associated with ribs (7). Herein, we present radiological and pathological findings of the second STABC case in the thoracic cavity in the literature and discuss the differential diagnosis.

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Submitted (Başvuru tarihi): 03.04.2016 **Accepted (Kabul tarihi):** 23.06.2016

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CASE

A 15-year-old boy presented with chest pain and dyspnea. Chest X-ray showed a homogenous mass in the right thoracic cavity. Thoracic computed tomography (CT) which was performed at another center showed a large predominantly cystic, well-defined mass (Figure 1). It had also multiple enhancing septations and irregular calcified areas. Magnetic resonance imaging (MRI) in our hospital demonstrated a lesion consisting of multiple cystic areas, the contents of which varied in intensity and the contours of which were lobulated. The lesion was located in the midline of the upper lobe of the left lung and about 77x68x70 mm in size with multiple septations. No marked diffusion limitation was observed. Following the injection of the contrast medium, contrast enhancement in the cystic walls and septa was observed. There was a remarkable atelectasis in the pulmonary parenchyma around the cyst. Bone structures and soft tissues of the chest wall were normal. The ribs were also normal. Surgical excision of the mass and pulmonary parenchyma by the side of mass was performed. Gross examination revealed a cystic lesion 80x70x70 mm in size with a hemorrhagic smooth surface. There were cystic structures filled with blood and the walls of which were calcific in the surfaces of the section. Microscopically, cystic areas lined by cuboidal cells were detected, with hemorrhaging in most of them. The septa were rich in fibroblasts and giant cells (Figures 2 and 3). There were also focal areas of calcification and osteoid (Figure 4). Hemorrhaging regions, inflammatory cells, and atelectasis in the pulmonary parenchyma adhering to the mass were also present. All of these pathological findings were consistent with those of an ABC. Clinical and pathological examinations during two-year-follow-up showed no abnormality or recurrence.

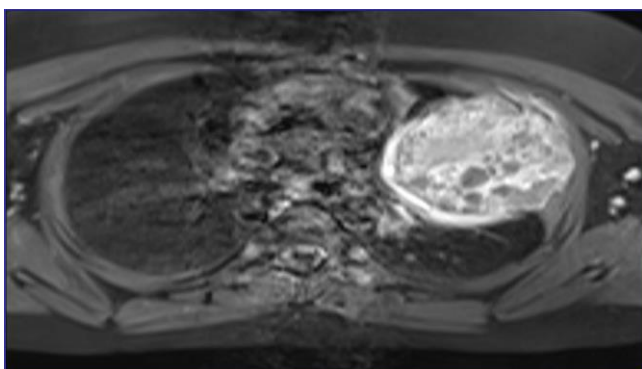


Figure 1: A computed tomography scan showing an irregularly shaped 77X68X70 mm mass

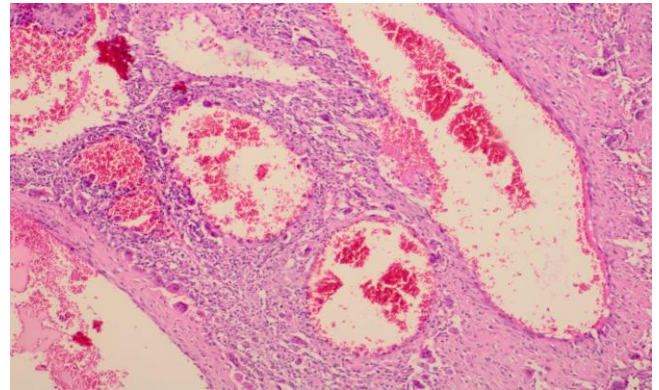


Figure 2: Hemorrhagic areas within a cystic space and multi-nucleate giant cells (H.E. X 200)

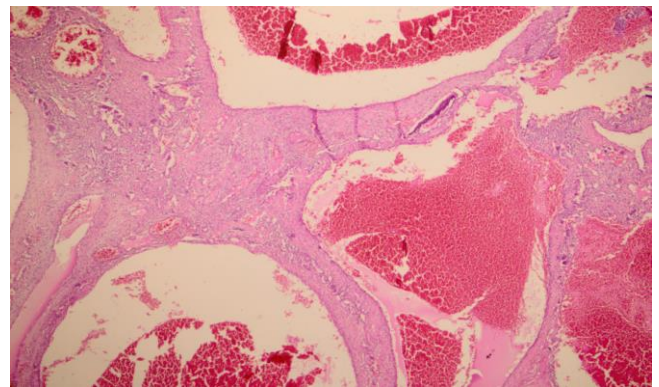


Figure 3: Hemorrhagic areas within a cystic space and multi-nucleate giant cells (H.E. X 200)

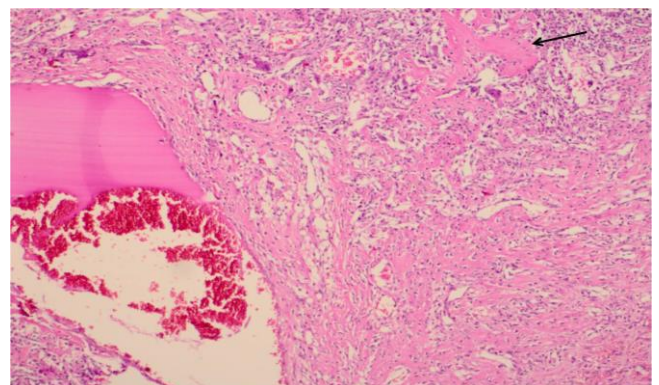


Figure 4: Osteoblastic rimming and osteoid element with aneurismal bone cyst (H.E. X 400)

DISCUSSION

Aneurysmal bone cysts were described as bone lesions until 1994; however, later cases of aneurysmal cysts of the soft tissue with ABC morphology on histopathological examination without bone involvement were reported. There were fewer than 20 well-documented cases of ABCs in the soft tissue in the literature until 2012 (3,8). Aneurysmal bone cysts can involve any bones, the most common site being end of the long tubular bones and vertebral column. The rib is a rare location of ABCs accounting for 2.7% of all cases (7,9). Although there have

been many cases of an intrathoracic ABC with rib involvement (7), there is only one case of an intrathoracic mass of the soft tissue without rib involvement (6,10).

Although the etiology of ABCs is unknown, circulatory disturbances and trauma have been proposed to be possible factors in the pathogenesis (7,11,12). The etiology of soft tissue ABCs is unknown, either. Nielsen et al. (2) suggested that a soft tissue ABC was a cystic form of myositis ossificans in that both had similar morphological features. However, it was reported in the literature that, unlike myositis ossificans, ABCs were neoplastic processes in that both the case presented by Nielsen et al. (2) and other cases reported had an abnormal karyotype. We believe that ABCs and myositis ossificans were distinct histological lesions. The most important diagnostic feature is provided by the maturation pattern characterized by a central cellular area, an intermediate zone of osteoid formation, and a peripheral shell of a highly organized bone in cases of myositis ossificans (2,13,14). This zonal pattern and bone maturation towards the peripherals of the lesion are not seen in cases of ABCs, which can be used in the differential diagnosis (15). Another lesion to be taken into consideration in the differential diagnosis of soft tissue and bone ABCs is a giant cell tumor. These tumors are associated with ABCs, since ABCs often contain a large number of giant cells. These two tumors can be mistaken for or can accompany each other. It is occasionally difficult to distinguish an ABC from a giant cell tumor, particularly, when a giant cell tumor exhibits bleeding, necrosis, and cystic changes. However, giant cell tumors usually occur in adults aged above 20 years, whereas ABCs are often found in the first two decades of life (16).

Extraskeletal telangiectatic osteosarcomas should be also distinguished from soft tissue ABCs. Histologically, atypical anaplastic cells may appear in osteosarcomas septa, while they are not seen in soft tissue ABCs. In the case presented here, there were bone trabeculae containing osteoblastic rims and osteoid; however, there was no anaplasia and cytological atypia (17). Due to its thoracic location, juvenile bone cyst, Ewing's sarcoma, eosinophilic granuloma, metastasis of neuroblastoma and leukemia, osteochondroma, callus tumor of the rib and chest wall hamartoma and all benign and malignant lesions of the rib must be kept in mind in the differential diagnosis of ABCs in children (9). All aforementioned lesions are associated with the rib; however, radiological imaging did not show an association of the lesion with the rib in the present case. The lesion was first diagnosed

as cyst hydatid in radiological examinations, since it was located in the thoracic cavity and the patient was a child. Besides, we suggested that it could be teratoma due to the young age of the patient. Histologically, the cystic spaces were filled with blood and the cyst walls were composed of bland spindle-shaped fibroblasts that grew in a fascicular or storiform pattern and were admixed with multi-nucleated osteoclast cells, hemosiderin-laden macrophages and trabecular bones. Osteoid was seen focally near the capsule (12,15).

Furthermore, CT and MRI are helpful in delineating the location and extent of the tumor and in identifying tumor tissues and local spread of a soft tissue mass. Soft tissue ABCs show a rather similar appearance to its osseous counterpart. Although radiological manifestations of benign and malignant chest wall tumors frequently overlap, differences in characteristic location and appearance occasionally allow a differential diagnosis to be made with confidence. Both aneurysmal bone cysts and giant cell tumors typically manifest as expansile osteolytic lesions and occasionally show fluid-fluid levels suggestive of the diagnosis. Juvenile bone cyst, Ewing's sarcoma, eosinophilic granuloma, osteochondroma, callus tumor of the rib, chest wall hamartoma, and leukemia are all benign and malignant lesions of the rib which must be kept in mind in the histopathological and radiological differential diagnosis of ABCs (9,18). Fluid-fluid levels can be seen in ABCs; however, this finding is not specific for ABCs. This appearance is also seen in other bone lesions and teratomas which contain areas of hemorrhage, cyst, or necrosis. Teratomas should be taken into consideration in the differential diagnosis of ABCs in children (19,20). The region where the study was conducted is a place in which cyst hydatid frequently appears, and it is most frequently seen in the lung and cyst hydatid located in the thoracic wall is also common (21).

An en-bloc resection with a clear margin of the lesion including the involved rib and soft tissue is the optimal treatment method for ABCs. They infrequently recur after marginal, but complete excision; therefore, this mode of therapy probably represents adequate treatment. About 90% of recurrences occur within two years (1,20). Fortunately, the case presented here did not have a recurrence during the three-year-follow-up period.

In conclusion, due to uncommon and extraordinary localization of soft tissue ABCs, a multidisciplinary approach with radiologists and pathologists should be followed for the diagnosis and differential diagnosis.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - T.C., F.B., N.B.; Planning and Design - T.C., F.B., N.B.; Supervision - T.C., F.B., N.B.; Funding - T.C.; Materials - T.C.; Data Collection and/or Processing - T.C.; Analysis and/or Interpretation - F.B.; Literature Review - N.B.; Writing - T.C.; Critical Review - T.C.

YAZAR KATKILARI

Fikir - T.C., F.B., N.B.; Tasarım ve Dizayn - T.C., F.B., N.B.; Denetleme - T.C., F.B., N.B.; Kaynaklar - T.C.; Malzemeler - T.C.; Veri Toplama ve/veya İşleme - T.C.; Analiz ve/veya Yorum - F.B.; Literatür Taraması - N.B.; Yazıyı Yazan - T.C.; Eleştirel İnceleme - T.C.

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