

Cases Report

Fatal Bilateral Congenital Mesenchymal Hamartoma of the Chest Wall

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The authors reported a newborn female baby, 36 weeks gestational age, 2,680 gms who developed respiratory distress at 2 hours after birth. Her chest roentgenograms showed a normal-sized heart, sparse lung markings and bilateral masses involving the posterior 6th to 8th ribs. Thoracotomy with partial removal of the left chest mass was performed when she was 12 hours age. Postoperatively, the baby developed progressive respiratory distress and expired at 29 hours – age.

Autopsy revealed bilateral nodular masses arising from the inner side of the posterior aspect of the 6th to 8th ribs, measuring 6 x 5 x 4 cm (right) and 7x 6 x 4 cm (left). The cut surfaces showed multicystic spaces containing blood. Histologically, many blood filled spaces with walls of fibroblasts, cartilage and bone tissue were noted. The diagnosis was mesenchymal hamartoma of the chest wall.

Mesenchymal hamartoma of the chest wall usually arises from the posterior or lateral portions of the rib and usually involves many ribs. Multi focal lesions and bilaterality are rare. The clinical presentation can be asymptomatic, mild or severe respiratory distress. Surgical resection is the treatment of choice.

Keywords: Mesenchymal hamartoma, Chest wall, Bilateral, Congenital, Fatal

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Mesenchymal hamartoma of the chest wall (MHCW) is very rare in infants. At least 77 cases have been reported⁽¹⁻³²⁾. The entity is usually present at birth or in early infancy with deformity of the chest wall and respiratory distress. This lesion is characterized by involvement of one or more adjoining ribs and it is composed of chondroid tissue, osseous tissue, immature mesenchyme and blood spaces. The behavior is that of a benign lesion. However, death occurring shortly after birth due to severe respiratory distress has been reported in twocases^(8,17).

The authors wish to report a fatal case of bilateral MHCW and believe that it is the first reported case in Thailand.

Case Report

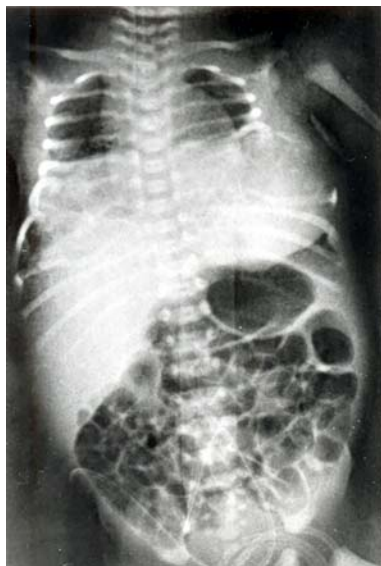
A 2,680 gram, female baby of 36 weeks gestation was noted to have dyspnea and retraction of the

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chest wall at 2 hours after birth. She was admitted to the Children's Hospital on September 8, 1983. Chest roentgenograms revealed a normal-sized heart, sparse lung markings and bilateral masses involving the posterior 6th to 8th ribs (Fig. 1). The masses caused ribs spreading and deformity. Thoracotomy with partial removal of the left chest mass was performed when she was 12 hours-age. Postoperatively, the baby developed progressive dyspnea and expired at 29 hours-age.

Pathology

Autopsy revealed bilateral nodular, well circumscribed masses arising from the inner side of the posterior aspect of the 6th to 8th ribs (Fig. 2). The left mass measured 7 x 6 x 4 cm and the right measured 6 x 5 x 4 cm. Both projected into the thoracic cavities and severely compressed both lungs. The cut surfaces of the masses revealed multicystic spaces containing blood, measuring up to 3 cm in diameter. The tissue surrounding the cysts consisted of grayish white soft tissue, cartilage and gritty bone tissue (Fig. 3).



A



B

Fig. 1 Radiographs reveal soft tissue masses involving 6th to 8th ribs both sides, masses cause ribs spreading and deformity, bone like density is seen in the masses, A: PA view, B: lateral view



Fig. 2 Gross pathology reveals masses in both sides



Fig. 3 Cut section of the mass reveals cystic cavities containing blood

Histologically, the tumors were composed of many blood filled spaces with walls of fibroblastic connective tissue (Fig. 4). In the solid areas, the tumors were composed of immature mesenchymal cells with transformation to cartilage that varies in size from small to large (Fig. 5). Enchondral ossification with bone marrow components resembling epiphysis was noted (Fig. 6).

Other autopsy findings showed marked bilateral pulmonary atelectasis and massive amniotic fluid aspiration. The right and left lungs weighed 20

and 17.3 gms, respectively. The brain showed anoxic change.

Discussion

Chest wall tumors in childhood include a wide range of benign and malignant lesions Mesenchymal hamartoma of the chest wall in infants is rare. To date about 77 cases have been reported⁽¹⁻³²⁾. The lesions were discovered at birth in about 55% of the cases⁽²²⁾. Four cases were diagnosed in utero with CT scan or ultrasonography^(8,14,19,20). In most cases, the lesions



Fig. 4 Microscopy reveals cavity containing blood, the wall consists of bone, cartilage and mesenchymal tissue (H&E X40)

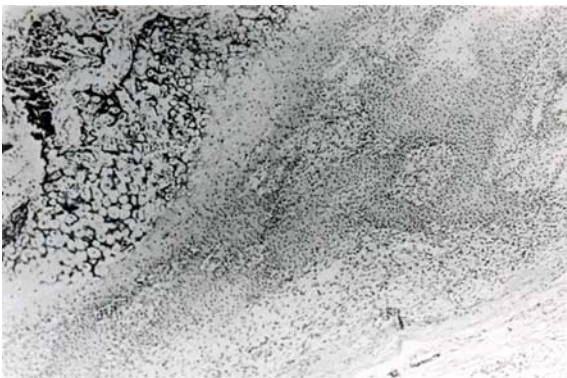


Fig. 5 Microscopy of the mass shows mesenchymal tissue, cartilage and bone (H&E X40)

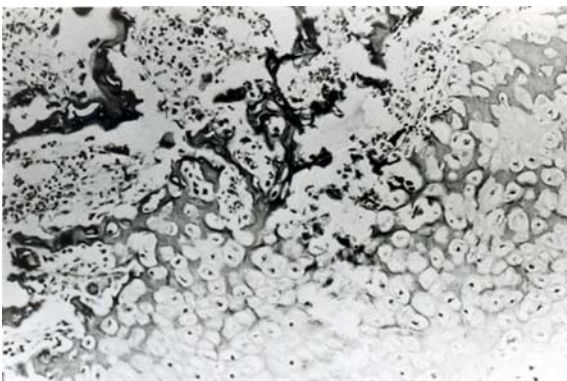


Fig. 6 Osteochondral-like junction with marrow tissue (H&E X40)

were detected during the first year of life⁽¹⁻³²⁾. Late diagnosis was reported in patients 4-13 years old and one adult female aged 26 years^(4,11,23,25). The male to female ratio is about 2:1^(17,22).

The mass usually arises from the posterior or lateral portions of the rib with projection into the thoracic cavity⁽¹⁸⁾. It is usually unifocal but multifocal lesions were reported^(16,23,25). Bilaterality was described in only four cases^(1,12,13,27). All were alive after resection. Many cases showed more than one rib involvement^(1-6,8-10,12-17).

The clinical presentation can be anything from mild respiratory symptoms, asymptomatic chest wall deformities to severe respiratory distress⁽¹⁻³²⁾. Fatal respiratory distress has been reported in two cases on their first day of life^(8,17).

The treatment of chest wall mesenchymal hamartoma is surgical resection of the tumor and portions of the involved ribs⁽¹⁸⁾. The two cases who received no surgical treatment were well after 16 years^(4,10). Chest wall deformity and severe scoliosis occurred after resection of the lesions^(3,8,10,13). Spontaneous regression was documented⁽²⁷⁾.

To the authors' knowledge, this patient is the first reported case of bilateral mesenchymal hamartoma of the chest wall in Thailand.

Conclusion

Bilaterality congenital hamartoma of the chest wall is extremely rare with about four reported cases. On autopsy of a 29 hours-age female baby disclosed bilateral mesenchymal hamartoma of the ribs with marked lung compression. The presented case appears to be the first reported case in Thailand.

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โรคมีเซ็นไคมีล ฮาร์มาโตมา ของกระดูกซี่โครงอกสองข้างตั้งแต่แรกเกิด และเป็นสาเหตุตาย

วิจิตรา เหมศรีชาติ, ปราณี เจริญขวัญ

รายงานผู้ป่วยเด็กหญิงแรกเกิด อายุครรภ์ 36 สัปดาห์ หนัก 2,680 กรัม เกิดภาวะหายใจลำบากเมื่ออายุ 2 ชั่วโมงหลังคลอด รังสีทรวงอก พบมีก้อนในทรวงอกทั้ง 2 ข้าง เกิดจากซี่โครงซี่ที่ 6-8 ด้านใน ด้านหลัง การผ่าตัดเอาก้อนบางส่วนด้านซ้ายออกเมื่อเด็กอายุครบ 12 ชั่วโมง เด็กไม่ดีขึ้นและตายเมื่ออายุ 29 ชั่วโมง

ผลการตรวจศพ พบก้อนใหญ่ขนาด 6 x 5 x 4 เซนติเมตร และ 7 x 6 x 4 เซนติเมตร ในช่องอกขวาและซ้ายตามลำดับ ก้อนเกิดจากด้านในด้านหลังของซี่โครงซี่ที่ 6-8 หน้าตัดเป็นช่องว่างมีเลือด ตรวจด้วยกล้องพบช่องว่างมีเลือดล้อมรอบด้วยไฟโบรบลาสต์ กระดูกอ่อน และกระดูก ให้การวินิจฉัยว่าเป็นมีเซ็นไคมีล ฮาร์มาโตมา ของทรวงอก

โรคนี้มีรายงานว่าก้อนมักเกิดจากด้านในของซี่โครง ทางด้านหลังหรือด้านข้าง มักพบว่าเกิดจากหลายซี่โครง ก่อนที่เกิดจากหลายซี่หรือ 2 ข้าง พบน้อยมาก คนไข้อาจไม่มีอาการ อาการน้อย หรือ อาการทางการหายใจลำบากมาก การรักษาโดยมากมักจะทำการผ่าตัดเอาก้อนออก มีน้อยรายมากที่ตายจากการหายใจลำบาก
