An infant with pericardial effusion

Tullu MS, Vaideeswar P*, Deshmukh CT, Lahiri KR, Pandit SP*, Chaturvedi RA*

Case Presentation

A six-week-old female child was admitted on 10th December 2003 with history of cough with cold for seven days and breathlessness for one day. The child was delivered at home and apparently had a low birth weight. The child had not cried after birth but did not receive expert neonatal care. She was un-immunized and was exclusively breast-fed. There was history of pulmonary tuberculosis in the father who had been treated for 2 years but his recent chest radiograph (April 2003) continued to show features of active tuberculosis. The chest radiographs of the mother and five-year-old sibling were normal.

On examination, the child weighed 2 kg, was afebrile with heart rate of 140/min and respiratory rate of 48/min with respiratory distress. Her blood pressure was 80/60 mm Hg. All peripheral pulses were well felt. Pallor, icterus, cyanosis, oedema and BCG scar were absent. The neck showed a soft, non-transilluminant, non-tender swelling 2 cm in diameter in the left anterior triangle. Systemic examination revealed muffled heart sounds. Respiratory system examination was normal. There was no hepato-splenomegaly. The child had mild hypertonia and brisk knee reflexes.

Investigations revealed haemoglobin of 11.6 gm/dl, leucocyte count of 17,500/ cmm (70% neutrophils and 30% lymphocytes) and platelets of 1.4 lakhs / cmm. Renal and liver functions were normal, so also was the blood gas examination. Blood groups of both the mother and the child were B positive. The chest radiograph revealed cardiomegaly and electrocardiogram showed low voltage complexes. Ultrasonography of the chest revealed pericardial effusion. This was confirmed on Colour Doppler, which had also indicated cardiac tamponade. The pericardial tap showed 27 ml sero-sanguineous fluid with 20,000 red cells/cmm, 500 polymorphonuclear leucocytes/cmm, 98 lymphocytes/ cmm, proteins of 243 mg% and gram-positive cocci in pairs on smear.

The child was treated with intravenous fluids, oxygen, intravenous ceftriaxone and two anti-tuberculous drugs (in view of the fact that the father was suffering from pulmonary tuberculosis). Colour Doppler after the pericardial tap revealed minimal residual effusion without tamponade and good biventricular function. In view of clinical worsening and respiratory distress, the patient was transferred to the Paediatric Intensive Care Unit (PICU) for close monitoring. The antibiotics were changed to vancomycin and gentamycin. He needed mechanical ventilation on the third admission day. A blood transfusion was given in view of anaemia (Haemoglobin- 7.6 gm%). The blood culture was negative. On the fourth day, the swelling in neck increased to diffuse soft swelling in sub-mandibular area and upper cervical region. On the fifth day, the child showed clinical improvement, was started on nasogastric feeds and weaned off the ventilator. Vancomycin was replaced by kloxacillin. Microbiologic investigations of the gastric lavage and pericardial fluid were negative. HIV testing of the mother was negative. On the sixth day of admission, a violaceous, 4 x 2 cm patch was noticed on the back with superficial ulceration. The respiratory system revealed bilateral crepitations. On the next day, right elbow skin discoloration was noticed. A computed tomographic (CT) scan of neck and chest (day 8) revealed subcutaneous oedema neck in neck, mild pericardial effusion, right middle lobe collapse with infiltrations in right middle and lower lobes. A repeat color Doppler showed thin rim of pericardial effusion. However the child had clinical deterioration with tachycardia, respiratory distress, thrombocytopenia and worsening of sensorium. He required re-ventilation and iotropnic support. Vancomycin was re-started. Blood culture continued to be negative. On 13th day, the neck swelling showed an increase in extent with brownish discoloration, clinically thought of to be cellulitis. CT scan of neck and chest, repeated on day 14 showed subcutaneous edema extending from neck to chest, laryngeal thickening, increased pericardial effusion (1.3 cm) and collapse /consolidation in lower lobes of lungs. A repeat pericardial tap performed on day 14 produced 30 ml haemorrhagic fluid. The child continued to deteriorate and sustained a terminal cardiac arrest on 17th day after admission.

Clinical Discussion and Differential Diagnosis

The working clinical diagnosis in the present case was myocar-
ditis, which is not a rare condition at this age. However, the color Doppler studies showed pericardial effusion with pericardial tamponade which occurs rarely as an isolated phenomenon at this age. At this age, transudative pericardial effusion is much more common and can be secondary to cardiac, renal, hepatic disorders, trauma or post-operatively following cardiac surgery. Exudative pericardial effusion is usually due to viral and other infections. Pyopericardium is rare, but can occasionally be seen in association with focus of bacterial infection elsewhere e.g. meningitis, pneumonia, septic arthritis and osteomyelitis. Initially the child was treated as a case of pyopericardium in view of a positive smear and leukocytosis. Though, there was history of tuberculous contact, the age, clinical presentation and the absence of acid-fast bacilli in pericardial tap fluid and gastric lavage samples were against the diagnosis of tuberculous pericardial effusion. The next likely possibility is of pericardial tumour. Most pericardial tumors present with cardiac tamponade with persistent or recurrent haemorrhagic pericardial fluid. Teratomas, mesothelioma, sarcoma, leukaemias and secondary tumors are the common types of tumours encountered. In view of the neck swelling, the skin lesion and CT scan findings, a possibility of mediastinitis was also considered. Later it was also noted that the patient’s response to antibiotics was poor. This finding along with a haemorrhagic pericardial fluid and unusual skin manifestations, a diagnosis of malignancy was strongly considered. However, this could not be confirmed by colour Doppler studies, pericardial fluid examination and CT scan.

**Autopsy Findings**

A complete autopsy was performed. Swelling of the neck and the two cutaneous discolourations were noted on general examination. In-situ examination of the thoracic cavity revealed distension of the pericardial cavity by fluid. The parietal pericardium was moderately thickened (Figure 1) with multiple, firm, slightly raised, well circumscribed, reddish lesions. Incision of the parietal pericardium revealed sero-sanguineous fluid. The inner surface of the pericardium was also studded with similar such plaques. The thymus was small in size and densely adherent to the base of the heart and could not be easily dissected, leaving behind firm congested tissue (Figure 1). The adventitia of the pulmonary trunk and ascending aorta were completely encased by similar, compressible congested tissue (Figure 1) and so were the superior vena cava and pulmonary veins. There were dense adhesions between the systemic veins, the pulmonary artery branches and the arch arteries to the trachea (Figure 2) due to firm congested soft tissue. Both the bronchi showed thickening of their walls and narrowing of the lumina due to dark, red tissue present along their entire lengths. Both atria were also covered by large red-brown plaques (Figure 3). Histologically, the plaques and the congested tissue described at the various sites, including the thymus and vascular adventitia were composed of multiple proliferating capillaries, some lined by plump endothelial cells (Figure 4). However, there was no evidence of malignancy. The lungs, which were firm and congested, revealed haemorrhage, bronchopneumonia and hyaline membrane formation. Haemangiomatous foci were not identified in the other organs, particularly the lungs and their pleurae, brain, liver and gastrointestinal tract. Also these changes did not reach up to the thoracic inlet. Thus a diagnosis of primary diffuse neonatal mediastinal haemangiomatosis was made.

**Discussion of the Condition**

Haemangiomas are a common-place, innocuous hamartomatous lesions involving the blood vessels, especially of the skin. Such cutaneous haemangiomas, in neonates and infants, are variably designated depending on the calibre of the vessel involved and the appearance. These are seldom of any clinical importance except when they are large or accompanied by complications. In rare instances, the haemangiomas acquire tremendous clinical significance, when they are multiple with multi-organ involvement or located at uncommon sites or associated with other anomalies. One such rarity is termed as neonatal haemangiomatosis.

Neonatal haemangiomatosis is an example of a non-heritable disorder characterized by the presence of multiple haemangiomas (multi-focal haemangiomatosis). In most instances, the disease follows a benign course, characterized by multiple cutaneous haemangiomas, with rare mucosal involvement and is designated as benign neonatal haemangiomatosis. The second more ominous subset is diffuse or disseminated neonatal haemangiomatosis (DNH), an example of which has been described in the case reported.

The first case of DNH described in the English literature was by Jaffe in 1929. Criteria for the diagnosis laid by Holder et al continue to be firmly entrenched and include recognition of visceral haemangiomas in the neonatal period, involvement of three or more organ systems and recognition of the benign morphology of these lesions. All the criteria were present in our case.

Liver is the commonest organ involved in DNH. Such infants present with a triad of hepatomegaly, congestive cardiac failure and anemia. The gastrointestinal tract and the central nervous system are the next organ systems. However, no organ has been spared by this disorder. A noteworthy feature in most cases is the constant presence of cutaneous haemangiomas with frequent occurrence in the oral, conjunctival or genital sites. Our case had many unique features. There were possibly only two cutaneous haemangiomas (as they were not biopsied during autopsy), a feature seen in only 10–15% cases of DNH. But, these lesions had evolved in the infantile period and exhibited rapid progression. Most interesting feature was a sharp restriction to the mediastinum with surprising sparing of the pleurae and the lungs. Such an occurrence has not been reported before, as also the diffuse affliction of the adventitial aspects of the arteries and veins, including the great vessels and exclusive bi-atrial involvement. The haemangiomatosis had also not extended beyond the thoracic inlet.

DNH is a life threatening disorder and the mortality rate (60-
95%) is mainly due to haemorrhage and other coagulation defects or cardiac failure. But, few cases have been reported in adults. In this patient, the breathlessness could be attributed to concomitant presence of pericardial effusion, lower lobe collapse possibly engineered by the proliferating capillaries in the wall of the bronchi, further complicated by the presence of severe bronchopneumonia. The present case also manifested with superior vena caval syndrome due to compression of the venous walls by proliferating capillaries, hindering the venous blood flow. The therapy would be multimodal including surgical resection, steroids, interferon, radiation with flash lamp pulsed dye laser.

References
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