Spontaneous bilateral chylothorax with fatal outcome in a patient with melorheostosis

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ABSTRACT

تم الإبلاغ عن حالة تعاني من تعظم النهايات المخطط متعدد البؤر لدى امرأة تبلغ من العمر 28 عاماً، في الذراع الأيسر، الصدر، العامود الفقري، وتصيب أيضاً النسيج الطري المتأثر. خضعت المريضة في السابق لعمليات جراحية وعائية متعددة. حضرت إلى المستشفى وهي تعاني من تكلس الصدر التلقائي على الجانبين. بعدما تلقت العلاج التحفظي وبدون نجاح توصلنا إلى وجود التهاب في الغشاء بجانب الرئة على الجانبين. ونتج عن ذلك إرجاع صاف للتسريبات على جانبي الرئة ولكن ازداد حالة المريضة الطبية سوءاً بشكل تلقائي نتيجة للترشيحات المتزايدة للنسيج الحشوي وزيادة التسريبات الجانبية للغشاء الرئوي داخل الفص. توفيت المريضة نتيجة لعدم كفاءة عمل الجهاز التنفسي بشكل عام. إن المرضى الذين يعانون من تعظم النهايات المخطط، قد ينتج عن إصابة النسيج الطري حالة مرضية المميزة، وفي أي مكان يحتمل حدوثها يجب أن تكون المعالجة تحفظية.

We report a case of progressive, multifocal melorheostosis in a 28-year-old woman, with involvement of the left arm, chest, spine, and impressive soft tissue involvement. In the past, she had undergone multiple vascular interventions. She presented with spontaneous massive bilateral chylothorax. After conservative treatment without success, we conducted bilateral pleurodesis. This resulted in a clear reduction of pleural effusions, but her medical condition subsequently worsened due to progressive parenchymatous infiltrates, and increased interlobal pleural effusions. She ultimately died of global respiratory insufficiency. In patients with melorheostosis, involvement of the soft tissue can result in distinctive morbidity, and whenever possible, treatment should be conservative.

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Chylothorax is a rare cause of pleural effusion. It has never been described in combination with melorheostosis, which is a progressive mesodermal disorder affecting both genders, characterized by hyperostosis of the cortical bone, and affecting soft tissue growth and development. Melorheostosis was first described by Léri and Joanny in 1922.[1] Since then, around 400 cases have been described in the literature.[2] The course is normally benign, and progression is slow. The minimum prevalence is reported to be 0.9 cases per million.[3] We describe a unique case of treatment-resistant chylothorax with fatal outcome in combination with progressive melorheostosis.

Case Report. A 28-year-old woman was hospitalized with a history of progressive orthopnea for 5 days. A multifocal melorheostosis of her left arm, including the clavicle, chest, and spine, as well as involvement of soft tissue, had been documented for over 3 decades since her birth. Her family history reveals no other members with melorheostosis. Soft tissue involvement included complex arteriovenous malformations, aneurysms and lymphangiectasis, predominantly of her left distal upper arm, mesentery, and thorax (**Figure 1**). Irregular cortical thickening of the left upper humerus was demonstrated on x-ray film (**Figure 2**). The periosteal bone formation, and the presence of a flowing candle-wax pattern are typical findings in melorheostosis. The grotesque vascular deformation of the left hand was illustrated by phlebography (**Figure 3**). She had undergone vascular surgery repeatedly, starting at the age of 2 years, followed by several embolizations, and surgical procedures. Clinical examination revealed labored breathing, with a respiratory rate of 24/minute, an oxygen saturation of 95% in room air, and decreased breath sounds mainly on the left side with dull percussion note. Her hearing, vision, and neurological assessments were without pathological findings. Her left upper limb was spastic and tenderly swollen, with discolored skin and distended veins. Chest x-rays on admission confirmed massive bilateral pleural effusions accentuated on the left side. The thoracocentesis evacuated 1.5 L of chyle with a triglyceride (TG) concentration of 16.8 mmol/L (TG

in serum 2 mmol/L). She was transferred to surgery for thoracoscopic ligature of the thoracic duct and bilateral tube thoracostomy. Initially, she was placed on a diet with oral medium chain TG, which was changed to total parenteral nutrition (TPN) from the eleventh day after admission, with the intent of minimizing chyle production. Total parenteral nutrition was continued for a total of 21 days. The average amount of fluid drained was 2800 ml per day, and neither ligature en masse of the thoracic duct nor TPN was able to reduce this quantity. Therefore, TPN was discontinued on day 32. Lymphography at this time showed massive reticular and cystic deformations of the mediastinal lymphatic system (**Figure** 4). A pleurodesis with talcum on the right side was performed 5 weeks after admission, and one month later a pleurodesis on the left side was performed. Chyle production decreased, allowing chest tubes to be removed, but her general condition did not improve, and the chest x-ray again showed a slight interlobar collection of fluid. Her physical condition worsened one month after the last operation. She developed global respiratory insufficiency, probably resulting from the new prominent parenchymatous infiltrates and increased interlobal pleural effusions due to fluid overload, chyle production, and probably combined with a beginning infection. She died on day 117 after being admitted to the hospital.

Discussion. Chylothorax is an infrequent cause of pleural effusion and is due to disruption of the thoracic duct, or one of its major tributaries. Approximately half of all chylothoraces are due to neoplasms (mainly lymphomas), resulting in obstruction of the duct, with subsequent leakage of chyle into the pleural space. Around 25% are traumatic, most commonly associated with cardiovascular surgery in which the left subclavian artery is mobilized. Direct thoracic trauma, subclavian vein thrombosis, and lymphangioleiomyomatosis can also lead to chylothorax. Approximately 15% of chylothoraces

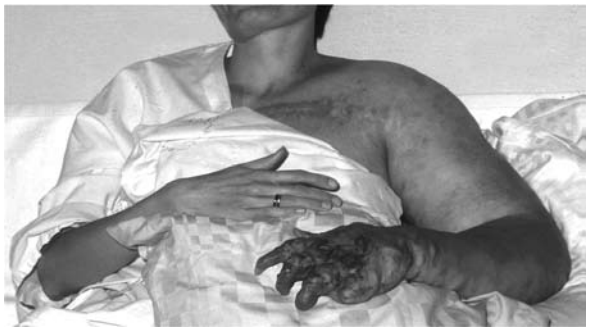

Figure 1 - Photograph on admission reveals the extent of soft tissue involvement with vascular deformities.

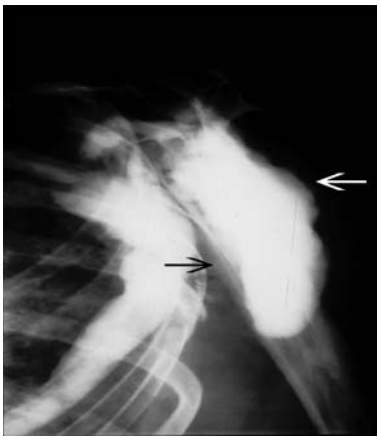

Figure 2 - Radiograph of the left upper humerus reveals distinct cortical thickening.

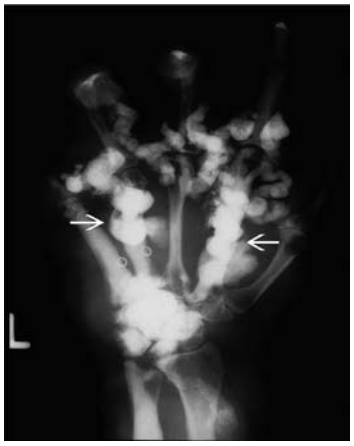

Figure 3 - Phlebography of the left hand shows massive vascular deformation and flexion contracture deformity of the fingers.

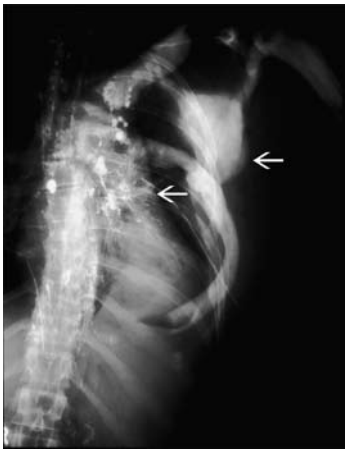

Figure 4 - Lymphography demonstrates extensive linear and globular dense cortical hyperostosis of the scapula, humerus, and the fifth rib. Massive reticular and cystic deformations of the mediastinal lymphatic system with multiple small leaks up to one cm in diameter. The thoracic duct cannot be identified.

are idiopathic.[4] To identify the cause and site of the lymphatic abnormality, patients with chylothorax should undergo lymphangiography.[4] We suppose that the cause of this chylothorax is the progression of melorheostosis. It is well known that this disease can be associated with soft tissue abnormalities of the affected limb, such as neurofibromatosis, skin pigmentation, linear scleroderma, hemangioma, arteriovenous malformations, arterial aneurysms, lymphangiectasia, lymphatic vesicles, and lymphedema. The differential diagnosis for other sclerosing bone dysphasias includes Paget's disease, von Recklinghausen's disease, osteopathia striata, and less common medical conditions like osteopoikilosis, Jaffe-Lichtenstein's polyostotic fibrous dysplasia, and others.[5]

In 1963 Morris et al,[6] reviewed 131 cases of melorheostosis. Many of them involved associated abnormalities of the affected limb, such as increased length, shortness, or deformity. In addition to the commonly noted muscle atrophy there were other abnormalities of the soft tissue of the affected limb, including scleroderma, lymphedema, hemangioma, and the like. In his review, 5 cases (4%) were associated with a lymphedema, and 7 (5%) showed vascular disorders, including arterio-venous malformations.

There is little experience with surgical treatment in patients with melorheostosis and vascular system involvement, and very little has been published on this topic. Treatment is usually symptomatic, and surgical correction of deformities to restore function is frequently pursued, but unfortunately, the postoperative course of these interventions is often complicated with vascular problems. We assume that in our patient, repetitive vascular surgery (7 embolizations and 6 subsequent surgical procedures) may have led to an aggravation of the vascular abnormalities. The initial treatment for chylothorax consists of chest tube drainage and dietary manipulations, in order to avoid dehydration, malnutrition, and immunological consequences. Somatostatin or the analog octreotide, seems to be a further valuable treatment option, which should be evaluated. If fluid loss exceeds 1.5 L/day for more than 5-7 days, or the leak persists for more than 2 weeks, conservative treatment should be abandoned and more aggressive treatment is then indicated, although this is associated with a higher risk of complications.[7,8] Some authors believe that an early operative intervention prevents nutritional demise.[9] In this situation, there are 3 main alternatives: 1) ligation of the thoracic duct, 2) pleurodesis, or 3) pleuroperitoneal shunt insertion. The thoracoscopic-assisted ligation of the supradiaphragmatic part of the thoracic duct is effective, and is usually not associated with long-term problems. In our patient, a bilateral pleurodesis was performed because of the unsuccessful prior medical and surgical approach, and the progressive deterioration of her nutritional condition. The amount of effusions decreased, but her general condition did not improve. Her physical condition worsened one month after the last operation. She developed a fatal global respiratory insufficiency probably secondary to the new prominent parenchymatous infiltrates, and increased interlobal pleural effusions. Cardiac failure or pulmonary embolism could be excluded. At this time she refused all further medical and surgical interventions. Respecting her wishes, an autopsy was not performed.

In our patient, several malformations of the lymphatic system with the progression of the underlying disease made a definitive surgical treatment of the bilateral chylothorax impossible. Chylothorax as a consequence of soft tissue involvement in connection with melorheostosis is very rare, and is a life-threatening disease. Our experience shows that in patients with melorheostosis and massive soft tissue involvement, multiple malformations of the lymphatic system correlate with the progression of melorheostosis, and are responsible for the failure of surgical treatment of bilateral chylothorax.

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