anticoagulated, a known cause of spontaneous thymic hemorrhage. However, there are many patients worldwide who undergo cardiac operations with subsequent anticoagulation, with few or no reports of spontaneous thymic hemorrhage. The risk associated between this phenomenon and anticoagulation is therefore not known.

Alternatively, patients with spontaneous thymic hemorrhage could have thymic disease predisposing them to bleeding, such as a thymic cyst. In our case, however, chest computed tomography was performed 6 years previously and was normal. Microscopically, a cystic wall did exist in conjunction with the hemorrhage. However, in neighboring normal thymic tissue hemorrhage was also seen, particularly in the medulla. For this reason, we conclude that this was a case of idiopathic spontaneous thymic hemorrhage in a normal thymus.

References


Recurrent Thymoma With Stiff-Person Syndrome and Pure Red Blood Cell Aplasia

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Stiff-person syndrome (formerly known as stiff-man syndrome) is a very rare autoimmune and neurogenic disorder, thought to present as a paraneoplastic variant in association with thymoma. Pure red blood cell aplasia is also a paraneoplastic disorder associated with thymoma. Although separate cases of stiff-person syndrome and pure red blood cell aplasia have been reported, we describe here what is to our knowledge the first case of recurrent thymoma with both stiff-person syndrome and pure red blood cell aplasia. We describe the successful treatment of the neurogenic symptoms of stiff-person syndrome and the progressive anemia associated with pure red blood cell aplasia by tumor excision.

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Thymoma with two synchronous paraneoplastic disorders is very rare. Resection of the recurrent thymoma may resolve the two paraneoplastic syndromes at once and alleviate the associated symptoms. Here, we report a case of recurrent thymoma with both paraneoplastic stiff-person syndrome (SPS) and pure red blood cell aplasia.

An 81-year-old woman with a growing recurrent mass on the left thoracic wall was referred to our department. She was experiencing slightly impaired ambulation caused by SPS and suffering dyspnea from progressive anemia, and she required frequent transfusions.

Thirteen years earlier, the patient had symptoms of muscle spasms of the axial and bilateral upper and lower extremities, lumbago resulting from deformities of the trunk caused by the spasm, and general asthenia. Computed tomography (CT) of the chest led to a diagnosis of thymoma, and an extended thymectomy was performed. The patient had no history of diabetes, epilepsy, or other neurologic diseases. However, the first day after the operation, she experienced severe pain in her bilateral legs and trunk on slight physical contact and cramping pain even in the absence of physical contact. Her condition deteriorated to the point at which she was unable to walk or maintain a sitting position unaided. The neurologist in our hospital examined her, and clinical laboratory tests revealed an elevated level of anti–glutamic acid decarboxylase (GAD) antibody; however, the patient was not diabetic. SPS was diagnosed, and glucocorticoids were administered. The patient then recovered gradually and was discharged from the hospital when she regained ambulatory function. She was followed up regularly by the neurologist.

In relation to the current admission, 13 years after thymectomy, CT of the chest revealed a recurrent chest wall mass, which had been slowly enlarging for 6 months (Fig 1). At this point, the patient presented with slightly progressive breathlessness and ambulatory disturbance, necessitating the use of a wheelchair. She had no other symptoms of SPS, for example, bilateral severe pain due to leg stiffness. Therefore, SPS was considered to be almost completely controlled with steroid administration (prednisolone, 10 mg/day).

Subsequently, the patient had progressive and refractory anemia, requiring frequent transfusion with concentrated red blood cells before the second operation. Her anemia was normocytic and normochromic, with a reduced percentage of reticulocytes. In addition, bone marrow aspiration revealed erythroid aplasia and a normal level of myelocytes. Her levels of serum iron, antiacetylcholine receptor antibody, antinuclear antibody, and immunoglobulin M against parvovirus B19 were all within normal limits, although the percentage of reticulocytes was decreased. At this stage, pure red blood cell aplasia, in addition to SPS, was diagnosed.

We decided to resect the tumor to control these paraneoplastic disorders. Preoperatively, we reduced the dose of steroid (prednisolone) to 5 mg every 2 days. Resection of the mass was achieved by the strategy of simultaneous chest wall resection and wedge partial resection of the left upper lung lobe, with the patient under general anesthesia combined with epidural anesthesia. The tumor was strongly fixed to the chest wall and the adjacent lung. We recognized that the tumor was invasive. However, adhesion of the other visceral pleura to the chest wall and mediastinum was small, so the operative procedure was achieved by video-assisted thoracotomy.

Postoperatively, the patient recovered and was discharged from the hospital with no major morbidity 2 weeks after the operation. Her progressive anemia had substantially improved. She needed transfusion only a few times for 6 months after the second operation, whereas she had previously needed transfusion once or twice a week for 6 months before the second operation. She was able to walk well using a stick 3 months after the operation.

On the basis of histologic examination, the tumor was classified as type B1 thymoma, predominantly a lymphocytic tumor, according to the World Health Organization criteria. The completely resected tumor had involved the lung and parietal pleura. A two-color flow cytometric examination revealed that 84.9% of all lymphocytes were CD4+ and CD8+, the typical phenotype of cortical thymocytes. After the second operation, the level of anti-GAD antibody in serum decreased to 13,000 U/mL (preoperative level, 19,000 U/mL; normal range, <1.5 U/mL).

Comment

Several kinds of disease are associated with thymoma, but thymoma with two synchronous paraneoplastic diseases is very rare, although the exact incidence is unknown. Resection of the recurrent thymoma may also resolve the paraneoplastic syndrome and alleviate the associated symptoms [1]. Here, we report a case of recurrent thymoma with both paraneoplastic SPS and pure red blood cell aplasia. To our knowledge, this is the first report of thymoma resection under these circumstances.

Thymoma is a slow-growing but malignant tumor, and surgical treatment to prolong survival should be adopted after careful consideration [2]. In particular, surgical treatment may not be suitable for elderly patients.
Bilateral Lung Transplantation in a Patient With Vascular Ehlers-Danlos Syndrome

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We describe the case of a 29-year-old woman with end-stage chronic obstructive pulmonary disease secondary to vascular Ehlers-Danlos syndrome. Because of critical deterioration, respiratory arrest, and complete lung failure, she required urgent implantation of a venovenous extracorporeal membrane oxygenator as a bridge to lung transplantation. After 6 days of extracorporeal life support, a successful bilateral sequential lung transplantation was performed. This is the first case of lung transplantation in a patient with a diagnosis of chronic obstructive pulmonary disease secondary to Ehlers-Danlos syndrome.

The vascular form of Ehlers-Danlos syndrome is a rare autosomal dominant disorder with an incidence of 1 in 10,000; it is characterized by joint hypermobility, skin hyperextensibility, characteristic facial features, and generalized connective tissue fragility, including easy bruising, thin skin with visible veins, and rupture of arteries, uterus, or intestines [1].

We report on a 29-year-old woman with vascular Ehlers-Danlos syndrome (vEDS), who deteriorated while on our waiting list for lung transplantation, eventually requiring extracorporeal life support (ECLS) bridging to lung transplantation.

At 12 years of her age, she presented with signs of early-onset emphysema of uncertain cause. A sweat test with negative results and electron microscopy for ciliary dyskinesia, immunoglobulin subtypes, and precipitants, and determination of autoantibodies and α1-antitrypsin level, did not reveal any abnormalities.

At age 26, computed tomography of the chest showed extensive emphysema, with no features suggesting conditions such as Langerhans cell histiocytosis or lymphangioleiomyomatosis. She was referred to a clinical geneticist with a suspicion of fragility of connective tissue, based on the patient’s background; at age 5 she had a congenital dislocating patella for which she had undergone two surgical procedures with tendon transfer. She also had irritable temporomandibular joint, and on examination she was of small dainty build, very slim, with very stretchy thin skin, and she had a joint hypermobility score of 2/9 involving her little fingers. Biochemical and molecular analyses showed reduction of type III collagen production and point mutation of the COL3A1 gene pertinent to vEDS.

Her clinical condition began to deteriorate, and 1 year later she was assessed and accepted on the waiting list for lung transplantation. Unfortunately, she continued to deteriorate clinically and required admission to our intensive care unit, with severe dyspnea, where she experienced respiratory arrest, requiring emergency intubation and mechanical ventilation. Despite our best efforts, complete lung failure developed, and it was decided to implant a venovenous extracorporeal membrane oxygenator system as a bridge to lung transplantation. The system was implanted percutaneously in a sedated patient and comprised a 27F Avalon Elite