phic with longitudinal or transverse ridging and loss of lunula and cuticles. Pleural effusions appear to be a later manifestation of the syndrome secondary to inadequate drainage by overstressed hypoplastic lymphatics rather than increased fluid production. The cause of bronchiectasis is unclear, but again, dysfunctional lymphatics are thought to play an important role with compromised drainage of secretions and local immune function.

Various malignancies have been associated with YNS, and one case of the yellow nails improved dramatically after resection of a laryngeal cancer. As in our case, Gupta et al reported similar improvement after surgery and chemotherapy for a carcinoma of the breast. Interestingly, improvement was seen in the fingernails only. Although partial or complete improvement in the nails may occur spontaneously in up to one third of patients, the temporal relationship and pace of the improvement strongly favors an association with successful treatment of malignancy. Possible explanations include direct involvement by tumor of already stressed and dysfunctional lymphatics or the elaboration of mediators such as peptide hormones that inhibit lymphatic function.

Various malignancies have been associated with YNS, and one case of the yellow nails improved dramatically after resection of a laryngeal cancer. As in our case, Gupta et al reported similar improvement after surgery and chemotherapy for a carcinoma of the breast. Interestingly, improvement was seen in the fingernails only. Although partial or complete improvement in the nails may occur spontaneously in up to one third of patients, the temporal relationship and pace of the improvement strongly favors an association with successful treatment of malignancy. Possible explanations include direct involvement by tumor of already stressed and dysfunctional lymphatics or the elaboration of mediators such as peptide hormones that inhibit lymphatic function.

Our case, we searched our pathology database for other soft-tissue tumors of the supraclavicular fossa and found no other case of sarcoma originating in this site. In addition, we performed a literature review of thoracic and neck liposarcomas to identify similar cases and discuss their clinical course.

**Myxoid Liposarcoma of the Supraclavicular Fossa***

Michael A. Morse, MD, Edward Bossen, MD; Thomas A. D’Amico, MD, FCCP, Warren Williamson, MD; and Richard Johnson, MD

Liposarcomas generally originate most often in the extremities or retroperitoneum, less frequently in the head and neck, and rarely in the thorax. We describe a particularly rare presentation of myxoid liposarcoma originating in the supraclavicular fossa. The mass was resected and has not recurred. We searched our pathology database for other soft-tissue tumors of the supraclavicular fossa and found no other case of sarcoma originating in this site. In addition, we performed a literature review of thoracic and neck liposarcomas to identify similar cases and discuss their clinical course.

**Key words:** myxoid liposarcoma; soft-tissue sarcomas; supraclavicular fossa

**REFERENCES**


Liposarcoma, the second most common soft-tissue sarcoma in adults, is separable into the following five major morphologic subtypes: well-differentiated, myxoid, round cell, dedifferentiated, and pleomorphic. The myxoid variant, which is characterized by the chromosomal translocation t(12;16) (q13;p11), is a lobulated soft-tissue tumor composed of slender, hyperchromatic spindle cells and signet-ring lipoblasts, a plexiform vascular pattern, and a myxoid matrix. Liposarcomas occur most often in the extremities and retroperitoneum, with only 5% occurring in the head and neck and 3% occurring in the thorax (primarily the mediastinum). Recently, we treated a patient with a myxoid liposarcoma that arose in a rarely described site, the supraclavicular fossa, and have reviewed our pathology database and the literature for similar cases.

**CASE REPORT**

A 71-year-old man presented with a history of several months of an enlarging, painless left supraclavicular fossa mass, measuring 7 cm. As a child, he underwent complete resection of a benign mass from his thigh. An excisional biopsy performed in March 1998 demonstrated a lobulated tumor, 5 cm in size, not arising from a lymph node. A histologic section (Fig 1) showed spindle-shaped cells and signet-ring lipoblasts in a myxoid matrix, which is consistent with myxoid liposarcoma. The surgical margins were involved with tumor, although no residual tumor was found at repeat resection. Postoperative staging studies demonstrated no residual mass in the supraclavicular fossa and no metastases in the remainder of the chest, abdomen, or bones. No adjuvant therapy has been given, and the patient has been observed without evidence of recurrence.

**DISCUSSION**

We searched our pathology database for biopsy specimens that were coded as being soft-tissue tumors and arising in the supraclavicular fossa, and we reviewed the available cases for which the final diagnosis was a soft-tissue sarcoma to identify other cases of liposarcomas. We found 19 other supraclavicular soft-tissue tumors that were not breast or lung cancer, lymphoma, adenocarcinoma of unknown primary origin, melanoma, or metastases from other known primary sites. The soft tissue-tumors were divided as follows: 13 lipomas; 1 lymph node metastasis of malignant fibrous histiocytoma originating in the scalp; 1 fibrosarcoma arising in the subclavian region in a patient who had undergone, as a neonate, resection of a hemangiendothelioma near the upper thoracic spine; 1 epithelioid hemangiendothelioma arising in the mediastinum and progressing locally into the subclavian region; 1 round-cell liposarcoma that occurred concurrently with a similar lesion in the liver and represented a metastasis from a primary calf tumor diagnosed 1 year previously; and 2 undifferentiated sarcomas that lacked clinical data.

A MEDLINE search was performed for supraclavicular, thoracic, or head and neck sarcomas to identify similar cases. Reports of supraclavicular sarcomas are indeed uncommon, compared with the more frequent finding of lipomas, which accounted for 13 of 20 mesenchymal masses resected from the supraclavicular fossa at our institution. Only two other reports (with four patients total) detailing the supraclavicular fossa as a primary site for liposarcomas have been published. Kindblom et al described one case each of well-differentiated round-cell liposarcoma and pleomorphic liposarcoma. All patients had recurrences requiring repeat resections or radiation therapy, and the patient with a pleomorphic liposarcoma died of multiple pulmonary metastases. Minic described a case of well-differentiated liposarcoma in the supraclavicular fossa that recurred despite resection and radiation therapy. In reviews of adult patients with a total of 1,305 head and neck soft-tissue sarcomas, only 30 patients (2%) had neck liposarcomas, although the exact location usually was not specified. Reviews of reports of thoracic liposarcomas demonstrate that most originate in the chest wall, mediastinum, or pleura, with no reports of a supraclavicular location. Myxoid liposarcoma is also notable for metastasizing to unusual sites, including the serosal surfaces of the pleura, mediastinum, pericardium, and diaphragm, and to extrapulmonary soft-tissue sites, including the peritoneum, chest wall, and breast. Lymph node metastases are extremely rare. We do not believe that our patient had a metastasis, although he had an unspecified tumor in his leg as a child 50 to 60 years earlier and recurrences after 30 years have been reported.

The prognosis for our patient is excellent. Factors that predict a better prognosis for liposarcomas are a well-differentiated and myxoid histology, a low percentage of the round-cell component, no spontaneous necrosis, a low num-

*From the Departments of Medicine (Dr. Morse), Thoracic Surgery (Dr. D’Amico), and Pathology (Dr. Bosson), Duke University Medical Center, Durham, NC; and the Departments of General Surgery (Dr. Williamson) and Pathology (Dr. Johnson), Southeastern Regional Medical Center, Lumberton, NC. Dr. Morse is a recipient of an American Society of Clinical Oncology Career Development Award and is supported by National Institutes of Health grant M01 RR00030. Manuscript received July 21, 1999; revision accepted October 15, 1999. Correspondence to: Michael A. Morse, MD, Duke University Medical Center, Box 2606, Durham, NC 27710; e-mail: m.morse@cg.duke.edu
Successful Pulmonary Thromboendarterectomy in a Patient With Klippel-Trenaunay Syndrome*

Beat Walder, MD; David P. Kapelanski, MD; William R. Auger, MD, FCCP, and Peter F. Fedullo, MD, FCCP

Klippel-Trenaunay syndrome (KTS) is a rare congenital disorder that consists of a triad of cutaneous vascular nevi, soft tissue or bony hypertrophy, and varicose veins or venous malformations involving one or more extremities. In addition to these main features, a wide range of associated conditions and complications involving the skeletal system, soft tissues, and cardiovascular system have been described. Among these complications is a propensity to thromboembolic events that, in a subset of patients, may lead to chronic thromboembolic pulmonary hypertension (CTEPH). CTEPH is a well-described entity; recognition of CTEPH is important because its natural history includes a high mortality rate and because it is potentially amenable to surgical intervention. This case report outlines the clinical history and management of a patient with CTEPH associated with KTS to underscore that evaluation for pulmonary thromboendarterectomy (PTE), a potentially curative procedure, should be considered in patients with KTS suffering from pulmonary hypertension.

**CASE REPORT**

A 24-year-old man with KTS was referred to University of California, San Diego Medical Center for evaluation of his pulmonary hypertension. The patient experienced a pulmonary embolic event 2.5 years prior to admission following a surgical procedure. There was no evidence of lower extremity venous thrombosis by duplex ultrasonography, although the examination was reported to be incomplete. He was treated with heparin, then warfarin for 4 months. He presented again 14 months later with dyspnea, chest tightness, and syncope. Ventilation/perfusion (V/Q) scan demonstrated multiple, segmental mismatched defects. Anticoagulation was restarted, and an inferior vena cava filter was placed. Three months later, a repeat V/Q scan was unchanged. CT confirmed the presence of intraluminal thrombus. Echocardiography revealed right atrial and right ventricular enlargement, with an estimated pulmonary artery systolic pressure of 75 mm Hg. He remained dyspneic with exertion (New York Heart Association class II), and was referred to University of California, San Diego Medical Center for further evaluation.

Physical examination was notable for a widely split S2 and a prominent pulmonic closure sound. Examination of the lower extremities revealed typical capillary malformations and hypertrophy of the left leg and right foot.

A V/Q scan revealed a global decrease in perfusion to the left lung, with additional defects involving the right upper and middle lobes; ventilation scan was normal. Echocardiography demonstrated moderate right atrial and right ventricular enlargement. The peak velocity of the tricuspid regurgitant envelope was 4.0 m/s, suggesting a peak pulmonary artery systolic pressure of approximately 75 mm Hg. The left popliteal to mid-superficial femoral veins could not be visualized by duplex ultrasonography. The remainder of the deep venous system of the lower extremity

**Key words:** Klippel-Trenaunay syndrome; pulmonary embolism; pulmonary hypertension; thromboendarterectomy; venous thromboembolism

**Abbreviations:** CTEPH = chronic thromboembolic pulmonary hypertension; KTS = Klippel-Trenaunay syndrome; PTE = pulmonary thromboendarterectomy; V/Q = ventilation-perfusion

---

*From the Divisions of Pulmonary and Critical Care Medicine (Drs. Walder, Auger, and Fedullo) and Cardiothoracic Surgery (Dr. Kapelanski), University of California, San Diego, School of Medicine, and UCSD Medical Center, San Diego, CA. Manuscript received July 26, 1999; revision accepted October 19, 1999. Correspondence to: Peter F. Fedullo, MD, FCCP, Division of Pulmonary and Critical Care Medicine, University of California, San Diego, School of Medicine, UCSD Medical Center, San Diego, CA; e-mail: bwalder@ucsd.edu