Case Report

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Surgical Eradication of Esophageal Carcinoma in a Patient with Klippel-Trenaunay Syndrome

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Key Words
esophageal tumor;
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Klippel-Trenaunay syndrome (KTS) is a rare congenital vascular disease of unknown etiology, and its occurrence with malignant tumor is extremely rare. We herein report a case of KTS presenting chronic gastrointestinal bleeding and complicating with adenosquamous cell carcinoma (ASCC) of the esophagus. The therapeutic dilemma of managing ASCC of the esophagus in a patient who combined with the rare congenital syndrome posed a difficult and interesting clinical problem. However, ASCC is also a very rare histological tumor in the esophagus, and its relationship to KTS has never been reported. In this article, we reviewed all the reported malignancies related to KTS. The possible surgical risks and complications, as well as preoperative imaging investigation, are also discussed.

KTS is a congenital disorder which is characterized by unilateral cutaneous hemangiomas, venous abnormalities, and bony and soft tissue hemihypertrophy. Similar to other congenital syndromes, KTS can be associated with various additional anomalies or diseases.[1,2] However, malignancy is usually thought to be an event independent from KTS. To date, only 5 cases of malignant tumors have been discussed regarding their possible relationship to KTS. Our patient had main features of this syndrome and developed unusual ASCC of the esophagus. Furthermore, we believe that surgical eradication of the malignant tumor may bring potential risks in this kind of patient, and stress that imaging studies are necessary before surgery.

CASE REPORT

A 64-year-old Chinese man presented with 1-month history of dysphagia to liquid diet and was admitted to our hospital for evaluation. Tracing back to the previous half year, he had already begun to have the symptom of poor appetite and loss of about 10 kilograms in the recent 3 months. Endosopic examination of the esophagus with tissue biopsy was performed before this admission, which demonstrated a protruding mass with irregular margin, about 5 cm in length, located in lower third of the esophagus. Malignant esophageal tumor is highly suspected. However, he denied any history of cigarette smoking or alcohol drinking, and his family history is also unremarkable. In the personal history was mention of congenital tortuous varicose veins over his right side trunk and upper extremity with ipsilateral soft tissue hypertrophy. These presenting features were evidence of Klippel-Trenaunay syndrome.

Barium swallowing study of the esophagus showed a lobulated filling defect with segmental mucosal irregularity, more than 5 cm in length, over the lower third of the esophagus, which extended downward to the esophagogastric junction. This finding was compatible with previous suspicion of esophageal carcinoma showing on endoscopic examination. Computed tomography (CT) with contrast enhancement was then planned for staging. Submucosal tumor over the lower third of the esophagus is observed on CT, and without evidence of apparent enlarged lymph node or metastatic

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lesion (Fig. 1). The preoperative imaging staging was $T_3N_0M_0$. In the meanwhile, previous tissue biopsy disclosed its malignant origin. Surgical resection was indicated to remove the apparently malignant esophageal tumor.

Tarry stool was noted during this admission, and Dipstick testing of stool revealed 4+ occult blood. Colonoscopy and upper gastrointestinal endoscopy, as well as red blood cell scan, were arranged. However, they did not demonstrate the exact site of gastrointestinal blooding. The preoperative laboratory studies also showed negative findings except mild decreased hemoglobin (Hgb 12.5 mg/dL) and hematocrit level. Venography was arranged due to the congenital hemodynamic malformation, and it revealed numerous superficial varicose veins over the patient right chest wall and upper limb with the absence of deep venous system (Fig. 2). Surgeons under deliberate surgical procedure performed successfully transhital esophagogastrectomy through abdominal midline incision with right-sided colon replacement. Mixed esophageal carcinoma with squamous and adenomatous differentiation was proved histopathologically. Postoperative staging was revised by pathologist as $T_3N_1M_0$ because of the evidence of local lymph node metastasis. The patient was then treated with radiotherapy and chemotherapy for the next 6 weeks.

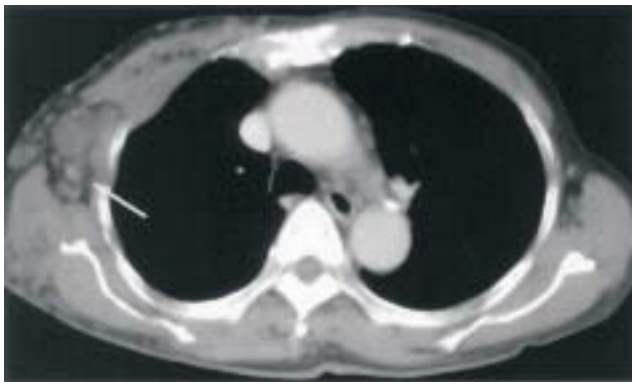

Fig. 1. Enhanced CT scan of the chest at the level of aortopulmonary window demonstrates multiple variable-sized soft-tissue masses (arrow) over the right side chest wall with right hemihypertrophy.

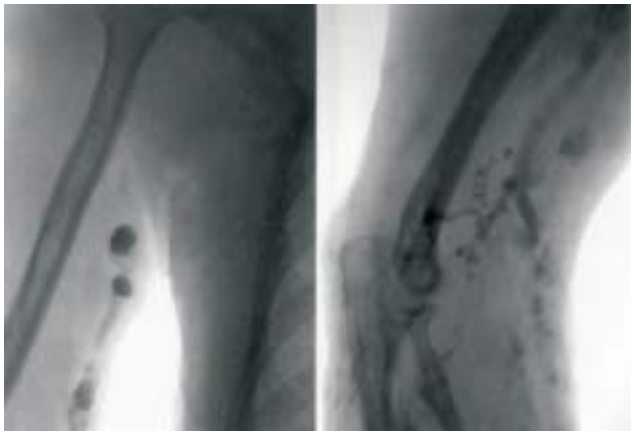

Fig. 2. Venography of the right upper extremity was performed, showing numerous superficial varicose veins over the patient right chest wall and upper limb. Absence of the deep venous system is also noted.

DISCUSSION

KTS is a rare, sporadic, congenital vascular disease, which may be due to disruption of normal mesodermal development in the early embryonic period. Unilateral cutaneous hemangiomas, varicose veins, and local gigantism with bony or soft tissue overgrowth are essential in making the diagnosis of KTS. Additionally, many associated anomalies have been correlated with this syndrome, such as visceral hemangiomas, lymphatic anomalies, and central nervous system abnormalities.[2-4] These associations reinforce the hypothesis that KTS may origin from mesodermal anomaly. Of special interest in our patient was the association of KTS with malignancy. To our best knowledge, only 5 reported malignancies have been described in regards to their association with KTS in the English literature.

Although the relationship of KTS with malignant tumors is controversial, it has still been suggested that this syndrome with some congenital defects may have more than a chance to develop malignancy.[5,6] Hemihypertrophy in patients with KTS is excessively associated not only with Wilms tumor, but also with nephroblastomatosis.[5,7] Chronic ulceration, a rare complication of KTS, may derive from congenital vascular malformation, and eventually developed cutaneous carcinoma in 1 reported case.[8] Our patient with KTS is the second to be reported with malignant esophageal cancer. Another similar case with this syndrome was a young female without evidence of potential nutritional or environmental risks.[9] Our patient

was, however, an old man but has no other predisposition to develop this type of tumor. This fact can perhaps lessen the possibility that the causes of esophageal tumor in association to KTS and suggests that KTS could be a causative factor in developing malignancy.

Surgical intervention is sometimes necessary in patients with KTS. However, the congenital vascular malformations in KTS can increase the risks of excessive bleeding or coagulation disturbance, resulting in a fatal condition.[10] Our patient was found to have the symptoms of unknown anemia and hematochezia. Angiodysplasia over the gastrointestinal tract should be considered in the differential diagnosis, especially in this kind of patient.[10,11] Although the current medical literature lacks well-designed studies of the various therapeutic modalities, several conclusions appear warranted. Diagnosis of the bleeding site during colonoscopy and the use of endoscopic hemostasis techniques may be necessary to control bleeding and prevent rebleeding.[11,12] Angiography is the most useful method to precisely demonstrate the wide range of the vascular malformations. Other imaging studies, like plain film, color Doppler scanning, CT, and even MRI, were much less sensitive to determine the presence of visceral hemangiomas.

In this report, we have reviewed all the reported KTS patients who were described in regards to their possible association with malignancies. Our patient is probably an additional case of KTS with malignant cancer, however, it was not possible to have direct evidence to prove. We also focus on the feasibility of surgical eradication of the ASCC of esophagus in patients complicating with this syndrome. When surgical intervention is considered in patients with KTS and unknown anemia, as in our present case, endoscopic evaluation of the gastrointestinal tract should be done. However, angiography is advised to delineate the possible visceral hemangiomas and give the surgeon more confidence to perform a successful operation.

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