marked improvement in the right-sided pressures, and a peak gradient of 8 mm Hg across the left ventricular outflow tract (Table 1). Aortic root angiogram showed moderate aortic regurgitation with prolapse of the posterior aortic cusp. Hydrogen arrival times showed absence of shunting. Chest x-ray film postoperaton is shown in Figure 2. Over the last three years he has remained free of chest pain and syncope.

DISCUSSION

Situs inversus totalis is a rare congenital anomaly, as demonstrated by mass radiographic screening in Norway which yielded an incidence of 1 in 10,000.4 Single coronary artery is also rare; a recent review suggested an incidence of 4 in 10,000.5 The chance of these two lesions occurring simultaneously is thus about 4 in 1 × 108. Hypertrophic subaortic stenosis is uncommonly associated with other cardiac anomalies.6 The constellation of defects in the above case is therefore unique.

In this patient, the unusually mild, long-term, clinical course is attributed to infundibular stenosis resulting in a natural “banding” of the pulmonary outflow tract. This is analogous to surgical banding of the pulmonary artery in order to prevent massive pulmonary blood flow and pulmonary hypertension in neonates deemed too young for closure of a ventricular septal defect.7

The weakness, chest pain, and syncope which developed in our patient at age 35 are attributed to the combination of left ventricular outflow obstruction and ventricular septal defect rather than to any effects of the single coronary artery which was widely patent angiographically. Complete surgical correction of the defects resulted in marked improvement in the hemodynamic and clinical states.

The development of aortic regurgitation after surgery was a result of closure of the high ventricular septal defect and was not present prior to surgery.

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Prophylactic Irradiation of the Lung in Ewing’s Sarcoma*

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A patient with Ewing’s sarcoma of the left scalula received prophylactic radiotherapy to the left lung following wide excision of the tumor. While the left lung remained free of tumor, there was metastatic invasion of the right lung within ten months of the initial diagnosis of the disease. Since Ewing’s sarcoma frequently metastasizes to the lungs, it is suggested that prophylactic irradiation of the lung may be of value in the prevention of metastases.

Ewing’s sarcoma, or diffuse endotheliona of bone,1 is an uncommon primary malignant neoplasm.2 The traditional treatment for patients with localized Ewing’s sarcoma has been either surgical or radiation therapy of the primary tumor.3 Controversy has existed as to the preferable method of treatment, but despite the high frequency of local control with both, cure is seldom effected because of rapid dissemination resulting from early hematogenous spread.4 5 The high probability of occult metastases being present at the time of diagnosis has stimulated the search for adjuvant systemic therapy designed to eradicate microscopic foci of metastases, namely, systemic chemotherapy6-8 or total-body irradiation.9 10

This report describes the effect of prophylactic irradiation of the lung in a patient with Ewing’s sarcoma.

CASE REPORT

A 26-year-old woman was diagnosed in January 1973 as having Ewing’s sarcoma. The primary site was the left scalula, with no evidence of distant metastases. Surgery, which included wide excision of the tumor and adjacent soft tissue, was followed by radiotherapy (Telecobalt90 unit). During March and April 1973, the patient received 3,000 rads (posteroanterior at a depth of 4 cm) to the left hemithorax (field size, 20 × 16 cm) in three weeks, and an additional 3,000 rads (posteroanterior at a depth of 4 cm) to the left scapular region (field size, 14 × 12 cm) in three weeks. Subsequently, by the end of April 1973, chemotherapy at dosages of 1.5 mg of vincristine per square meter of body surface area and 400 mg of cyclophosphamide per square meter of body surface area was given once weekly for six weeks and then once biweekly. A chest x-ray film in July 1973 was normal (Fig 1).

In September 1973, transient signs and symptoms of radiation-induced pneumonitis were noted. Ten months after surgery (November 1973), a small single metastatic lesion appeared in the right lung, and despite further intensive chemotherapy with adriamycin (30 mg/sq m of body surface area) and vincristine (1.5 mg/sq m of body surface area), an increase in the size and number of lesions in this lung could be seen on a repeat chest x-ray film in July 1974 (Fig 2).

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Skeletal metastases were first demonstrated in May 1974, and palliative irradiation to various bone metastases was given. Further irradiation of the tumor masses in the right lung with 2,700 rads in two weeks, anteroposterior and posteroanterior (field size, 15 × 10 cm), failed to bring about any improvement (September 1974, Fig 3). Additional chemotherapy, including bischloroethylnitrosourea, dactinomycin (actinomycin D), and steroids, was given for five weeks. The patient's condition further deteriorated with the occurrence of cerebral metastases. She died in October 1974 because of septic shock. Permission for autopsy was refused.

**Figure 1.** Normal chest radiograph (July 1973).

**Figure 2.** Metastases in the lower right lung (July 1974).

**Figure 3.** Diffuse metastases in the right lung (September 1974).

Low-dose severe radiation-induced pneumonitis following prophylactic pulmonary irradiation has recently been described in conjunction with previous treatment with dactinomycin.16

In order to attain control of this highly malignant tumor, our patient was irradiated through the whole left hemithorax, with an additional dose to the left scapular region. Metastases appeared in the right lung, which had not been prophylactically irradiated; however, the left lung, which had been prophylactically irradiated, showed no radiologic evidence of tumor spread throughout the entire course of the disease. Intensive therapeutic irradiation of the metastases in the right lung was without response.

The recent suggestion of prophylactic whole-body irradiation in Ewing's sarcoma5,10 has not gained general acceptance. Yet our clinical observation, with one lung prophylactically irradiated and the other acting as a control, suggests that prophylactic pulmonary irradiation may have a place in the combined regimen of radiotherapy and chemotherapy for patients with Ewing's sarcoma.5,8,15

**References**


**Discussion**

Metastases in Ewing's sarcoma occur in 75 to 85 percent of cases within two years after diagnosis.11,12 Local recurrence after radiotherapy has been described in 2712 to 38 percent14 of patients. The lungs were the first site of metastases in 57 percent (34) of a series of 59 patients and at autopsy were involved in 14 of 18 patients.15 These findings provide the rationales for prophylactic pulmonary irradiation in Ewing's sarcoma, and the doses employed are generally well tolerated.

Pulmonary Varices*
Report of Two Cases and Review of the Literature

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Two cases of pulmonary varices are presented. The lesion is rare, and only 32 cases have been reported in the literature. Although the lesion is benign in nature, complications are possible, and death has occurred from rupture of the varix intrapleurally or into the bronchial tree.

The aneurysmal dilation of a pulmonary vein, known as pulmonary varix, is a relatively rare lesion and should be distinguished from the more common arteriovenous malformation. According to Rizk and associates,1 the first case of pulmonary varix was reported by Puchelt in 1843. Since then, only 32 cases have been recorded.3-21

Herein we report two cases of our own and review the pertinent literature.

Case Reports

Case 1

A 45-year-old asymptomatic man was referred because of a

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FIGURE 1. Plain chest x-ray film showing well-delineated mass (arrows) in lower part of right hemithorax.

round shadow in the right lung which was discovered on routine x-ray examination of the chest. The lesion was 2 cm in diameter and located in the middle lobe. Findings from routine examinations were normal. At thoracotomy, the lung looked normal, and no palpable mass could be found. The chest was closed, and the chest x-ray film taken immediately after surgery showed the round shadow still to be present. The chest was reentered; and, again, careful palpation failed to reveal any mass. The middle lobe was resected, and the pathologic examination revealed a pulmonary varix. The patient had an uneventful recovery.

Case 2

A 18-year-old woman was recently referred because of a well-delineated shadow discovered incidentally at x-ray examination of the chest. The mass was located in the medial aspect of the lower lobe, close to the hilum and pericardium, extending down to the diaphragm (Fig 1). A vascular malformation of the lung was suspected, and a pulmonary angiographic examination was performed, confirming the

FIGURE 2. Angiocardiogram (early arterial phase). Mass is not yet opacified.

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