been described, and oxygen administration may be dangerous in these patients.1,3,4

Noisy respiration, somnolence, cyanosis, congestive heart failure, and signs of pulmonary artery hypertension were the presenting features in this child with congenital ankylosis of the temporomandibular joint. The congenital anomaly produced respiratory and feeding difficulties aggravated during intercurrent respiratory infections and suggested the diagnosis of cor pulmonale secondary to upper airway obstruction. The cardiac catheterization data and the rapid improvement following tracheostomy and bilateral condylectomy confirmed the clinical impression. To the best of our knowledge, this is the first description of congenital ankylosis of the temporomandibular joint as a cause of chronic upper airway obstruction and cor pulmonale which can be successfully treated.

REFERENCES

Tuberculous Aneurysms of the Descending Thoracic Aorta*

Report of a Case with Fatal Rupture

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Tuberculous aortic aneurysm is a rare disease entity. The majority of affected patients succumb to perforation and exsanguination. The only chance for survival and cure is by resection and prolonged antituberculosis chemotherapy. Our case illustrates the high risk of rupture of tuberculous aortic aneurysms. Post-mortem examination revealed that the mechanism of aneurysm formation was by direct caseous involvement of the descending thoracic aorta from a juxtaposed left upper lobe parenchymal tuberculous process. Our findings also favor the concept that miliary dissemination (in the presence of tuberculous aortic aneurysm) is the result rather than the cause of the tuberculous aortic process.

Over one hundred cases of tuberculosis involving the aorta have been reported in the literature. Almost one half of these cases were associated with aneurysm formation, while the other one half were examples of tuberculous aortitis without aneurysm formation. The incidence of thoracic and abdominal aortic involvement is about equal.2 Very rarely is the ascending aorta involved.8 Up until the present time, only 13 cases of tuberculous aortic aneurysms have undergone resection with seven considered cured of the disease.1,4

Recently, we have encountered our first case of tuberculous aortic aneurysm. However, it was unfortunate that rupture and death occurred about 48 hours prior to the planned resection.

CASE REPORT

A 49-year-old black female schoolteacher was admitted on Jan 17, 1978, with a 2% month history of weight loss, cough with minimal sputum production, and shortness of breath. These symptoms had progressively become worse to the point that she had been confined to bed for several days because of extreme weakness and shortness of breath. As a schoolteacher for the previous ten years, she had yearly chest roentgenograms which were reportedly normal. Tuberculin skin tests were periodically done and were negative. In May 1977, she was told her tuberculin skin test was positive, and her chest x-ray film was interpreted as abnormal with an ill-defined infiltrate in her left lower lobe. She was advised to start on antituberculosis therapy, which she refused and she was subsequently lost to follow-up. She recalled that she was doing well during this time until November 1977, when she began feeling ill and losing weight. By late December, she was having night sweats, nonproductive cough, generalized weakness, and shortness of breath. She consulted her family

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physician who prescribed antibiotics and told her she had mild pneumonia, although no chest roentgenogram was done. Since that time, she had progressively deteriorated until her admission. She lost a total of 15.5 kg (60 lb) in a period of 2½ months.

On admission, she appeared ill, debilitated, and markedly short of breath. Vital signs were as follows: blood pressure, 136/80 mm Hg; pulse rate, 140 beats per minute; respiration, 32 per minute; and temperature, 37°C.

On physical examination, the lungs had diffuse dry rales without dullness or rhonchi. Abdominal examination revealed a slightly enlarged liver. Pitting edema of both lower extremities was noted.

Laboratory examinations were unremarkable except for serum albumin value of 2.6 gm/100 ml and arterial blood gas levels of pH 7.55, Pco₂ of 32 mm Hg, and Po₂ of 48 mm Hg.

Admission chest roentgenogram (Fig 1) showed diffuse, coarse reticulonodular infiltrates throughout both lung fields which appeared to be more pronounced in both apices, consistent with far-advanced pulmonary tuberculosis.

Course in the Hospital

About eight hours following admission, the patient suffered respiratory arrest. She was resuscitated successfully but required ventilatory support. Tracheobronchial washings and sputum examination showed acid-fast organisms. Regimens of isoniazid, ethambutol, and streptomycin were started.

On Jan 30, 1978, after almost two weeks of ventilatory support, she was weaned successfully from the respirator. She continued to show good progress with clearing of lungs as seen on her chest roentgenogram. Lumbar puncture revealed normal pressures and clear fluid was negative on smear and culture for acid-fast bacillus. Repeat sputum and gastric aspirates examination showed acid-fast bacillus. Her tuberculin skin test was positive.

In the next several weeks, she had progressive clinical improvement, with clearing of the diffuse pulmonary process.

**Figure 1.** Changes are consistent with far-advanced pulmonary tuberculosis.

**Figure 2.** Arteriography confirms presence of saccular aneurysm of proximal descending thoracic aorta.

With clearing evident on x-ray film, an abnormal prominence over the aortic knob became more pronounced, and the diagnosis of tuberculous aortic aneurysm was entertained. Aortography was performed and confirmed the presence of a saccular descending thoracic aortic aneurysm (Fig 2).

She was then scheduled to undergo resection of the aortic aneurysm. While preparation for surgery was in progress, she suddenly developed generalized seizures, massive hemothorax, and finally cardiopulmonary arrest. Resuscitation failed,

**Figure 3.** Medial view of left lung showing area of rupture of false thoracic aortic aneurysm into juxtaposed posteromedial aspect of left upper lobe where tuberculous caseous necrosis is evident (black arrow). Groove created by descending thoracic aorta is also evident (white arrow). Dark area at center is site of hemorrhage as aneurysm leaked out into pleural cavity.
and the patient died.
Post-mortem examination was done.

POST-MORTEM FINDINGS
Pathologic examination revealed an active tuberculous process in both lungs with identifiable acid-fast bacilli. On the posteromedial aspect of the left upper lobe was a 4 x 3 x 0.5 cm soft, whitish-yellow friable tissue mass (Fig 3). This was juxtaposed with the anterior aspect of the proximal descending thoracic aorta about 2.5 cm distal to the origin of the left subclavian artery. On dissection, it was found that in the center of the mass was a "channel" which communicated with the aortic lumen on one end and left upper lobe bronchus on the other. The "hole" in the aortic wall was about 1 cm in diameter. It was rounded with smooth edges. There was no evidence of an old traumatic tear, arteriosclerotic plaque, or dissection.

These findings explained the terminal events of this patient. Significant hemoptysis was precipitated by the rupture of the aneurysm into the lung parenchyma and left upper lobe bronchus, as was further confirmed by the large amount of clotted blood in the tracheobronchial tree. On the anterior aspect of the left upper lobe mass, which was a false aneurysm, leakage into the free pleural space was apparent, which explained the presence of almost a liter of clotted blood in the left pleural cavity.

Although no microscopic examination was performed, the remainder of the aorta was normal by gross examination.

DISCUSSION
Tuberculous aneurysm of the aorta is a rare disease entity. There are approximately 50 cases reported in the literature, with a few over 50 more cases of tuberculosis reported involving the aorta without aneurysm formation.1 The majority of this latter group of patients was diagnosed on post-mortem examination. Similarly, among those with aneurysm formation, the majority succumbed to rupture and exsanguination; hence, most diagnoses were also made postmortem. To our knowledge, there are only some 13 cases of tuberculous aortic aneurysm resection reported in the literature with nine survivors, although two died at 13 and 19 months following surgery with complications related to tuberculosis.1 It is also agreed by most that the only chance of survival in this group of patients is early surgical resection combined with preoperative and prolonged postoperative antituberculosis chemotherapy.1,3,6 The high propensity for rupture of this type of aneurysm is better understood by the knowledge of the mechanism of their development.

Almost all tuberculous aortic aneurysms are not true aneurysms. There are a few reported cases of dissecting aneurysm secondary to tuberculous aortitis.6 Most of the authors who have some experience with this problem agreed that in the majority of cases, the aortic wall involvement by the tuberculous process, is merely an extension of a primary caseous process involving the lungs, periaortic lymph nodes, vertebra (Potts' disease), pericardium, or pleura.4,5 In only a few cases, a true hematogenous spread of bacteria into the aortic intima or vasa vasorum has been implicated. However, the absence of reports of tuberculous aortic aneurysm among large reviews of miliary tuberculosis7 highly suggest that systemic bacterial dissemination is rarely, if ever, the cause of the aortic tuberculous process. In essence, most tuberculous aortic aneurysms are false aneurysms representing a transmural perforation of the aortic wall by a tuberculous process that has been walled-off with blood clot.

Almost all patients with tuberculous aortitis have tuberculous lesions elsewhere in the body, most often in the lungs. Some of the reported cases have patterns of miliary tuberculosis or chest roentgenogram.2,4,6 The relationship between tuberculous aortic aneurysm and miliary tuberculosis has been debated. However, the majority believes that the miliary process in the presence of tuberculous aortic aneurysm is the result of intravascular dissemination of organisms originating from the aortic tuberculous process.4,5 We believe that a similar mechanism was involved in our case.

Knowing the dangerous nature of this process, it appears that immediate resection is the only way to improve survival. Prolonged postoperative antituberculosis chemotherapy should be utilized. It is probably reasonable to urge, despite the inanition and debility usually seen in such patients, that "the patient is too sick not to operate. During surgery, the aneurysm should be approached with great caution as with any false aneurysm. A difficult resection should be anticipated because of expected extensive adhesions and granulation tissue formation. A double-lumen endotracheal tube should be used if possible, and proximal and distal control of the aneurysm should be obtained before actual manipulation or dissection of the aneurysm is started. Finally, these patients should be followed-up closely for several years for possible reactivation of the tuberculous process in the area of resection or the development of similar process in other vessels as has been reported.1

REFERENCES
6 Meehan JJ, Faster BH, Torre AV: Dissecting aneurysm of the aorta secondary to tuberculous aortitis. Circulation 16:615-620, 1957

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