Ortner Syndrome with Recurrent Pericardial Effusion:
A Diagnostic and Therapeutic Dilemma
AKM Monwarul Islam, Muhammad Toufiqur Rahman and Mahboob Ali

ABSTRACT
Ortner syndrome is a rare condition characterized by hoarseness of voice in association with a cardiovascular disease. It is caused by compression of the left recurrent laryngeal nerve by the pulmonary artery or left atrium. Mitral stenosis is a well-recognized cause, however, a number of cardiac and non-cardiac conditions have also been described. Prognosis of Ortner syndrome depends on the underlying aetiology as well as the duration of illness. The case presented here describes a 35-year-old man with hoarseness of voice with recurrent pericardial effusion. Initially, a microbiologically proven diagnosis of tuberculous aetiology with resistance to Rifampicin was made; lack of optimum response and recurrence of pericardial effusion lead to subsequent diagnosis of metastatic adenocarcinoma. The patient responded to some extent to systemic and intrapericardial chemotherapy. Immunocompromized state associated with malignancy may predispose to infection including tuberculosis.


INTRODUCTION
Ortner syndrome or the cardiovocal syndrome is a rare condition characterized by hoarseness of voice due to left recurrent laryngeal nerve palsy in association with a cardiovascular pathology. Left recurrent laryngeal nerve palsy brings about the hoarseness. Mitral stenosis is a common cause, but a myriad of other cardiac and non-cardiac conditions may be responsible as well. The case presented here describes Ortner syndrome in association with recurrent massive pericardial effusion.

CASE REPORT
A 35-year-old non-smoker, normotensive man presented with low-grade fever, cough, occasional haemoptysis for 3 months and hoarseness of voice for 1 month. Physical examination was unremarkable. His total leucocytic count was 9,700/mm³, ESR was 110 mm in first hour and Mantoux test was positive (15 mm). Chest X-ray revealed left hilar lesion. Fiberoptic bronchoscopy found left vocal cord palsy (Figure 1). Acid-fast bacilli were found in bronchoalveolar lavage fluid in both Ziehl-Neelsen staining and culture in Lowenstein-Jensen media. He was offered anti-TB drugs with standard regimen, but the response was poor. While on anti-TB therapy, within 1 month, the patient developed progressive breathlessness. Chest X-ray showed features of pericardial effusion i.e. increased transverse diameter of cardiac shadow with penciled outline and oligemic lung fields (Figure 2). ECG revealed low QRS voltage i.e. < 5 mm in limb leads and < 10 mm in precordial leads along with sinus tachycardia (Figure 3). There was huge pericardial effusion (30 mm highest collection) on echocardiography. Pericardiocentesis was done, the fluid was exudative. Spiral CT scan of chest revealed sub-carinal and left hilar lymphadenopathy and bilateral pleural effusion. Meanwhile, the sensitivity report of the cultured TB bacilli was available and revealed resistance to rifampicin, but sensitivity to INH, ethambutol and streptomycin. A diagnosis of drug-resistant tuberculosis, huge pericardial effusion and Ortner syndrome was made. The patient was treated conservatively with INH, ethambutol, pyrazinamide, streptomycin and ofloxacin. Over the next months, there was little improvement, pericardial fluid re-accumulated, and the hoarseness of voice did not improve significantly. The patient was counseled regarding the difficulties in treating the disease, possible course, need for second-line anti-TB drugs and their possible side effects. Meanwhile, left-sided pleural effusion developed, pleural biopsy abroad reported presence of suspicious malignant cells. Fine-needle aspiration cytology (FNAC) of cervical lymph nodes established the diagnosis of metastatic adenocarcinoma (Figure 4). Subsequent search failed to localize the primary lesion. Repeat investigations for tuberculosis including adenosine deaminase activity, AFB staining and culture for acid-fast bacilli were negative. The patient was offered 6 cycles of palliative chemotherapy. Cisplatin was instilled intrapericardially and intrapleurally to prevent recurrence. General condition of the patient improved significantly, and there

660 Journal of the College of Physicians and Surgeons Pakistan 2013, Vol. 23 (9): 660-662
were almost no fluid in the pericardial and pleural spaces in follow-up.

The patient was counseled regarding the difficulties in treating the disease, possible course and prognosis. All measures taken were for palliation, to reduce morbidity and to prolong survival. Since the first medical contact for the present illness, one and a half years has passed, and the patient is alive 3 months after completion of 6 cycles of chemotherapy and intrapericardial cisplatin instillation.

**DISCUSSION**

Ortner syndrome has been described in association with a number of cardiac and non-cardiac conditions including mitral valvular disease.\(^1\)\(^-\)\(^4\) The basic abnormality is paralysis of the left recurrent laryngeal nerve, most commonly, by the pressure in the pulmonary artery.\(^4\) However, other mediastinal structures may cause the compression, or the nerve may be injured by mechanical or thermal injury. In the present case, the left recurrent laryngeal nerve was injured most probably by pressure over the pulmonary artery caused by huge pericardial effusion, and not due to involvement by the malignancy itself, as because the Ortner syndrome was evident long before the appearance of lymphadenopathy.

Both tuberculosis and malignancy are common causes of pericardial effusion, the former is more important in developing countries while the latter in developed world.\(^5\)\(^-\)\(^7\) However, multiple pathologies contributing to the aetiopathogenesis of pericardial effusion has been reported only rarely. *Salmonella enteritidis* was cultured from the pericardial fluid of a 64-year-old immunocompetent lady with untreated hypothyroidism, who presented with massive pericardial effusion.\(^8\) In the present case, tuberculous aetiology of pericardial effusion appears to be certain as culture of *Mycobacterium tuberculosis* from pericardial fluid or tissue is enough for diagnosis of tuberculous aetiology of pericardial effusion.\(^9\) Metastatic adenocarcinoma probably caused the pericardial effusion, and the associated immunocompromized state might favour new infection or activation of a previous infection with *Mycobacterium tuberculosis*.

Strikingly, the tubercular bacilli were resistant to one of the most useful first-line anti-tubercular drugs, namely, rifampicin. This is not unusual in this era of multi-drug resistant (MDR) and extensively-drug-resistant (XDR) tuberculosis. The malignancy and drug-resistant TB may

---

**Figure 1:** Fiberoptic bronchoscopy shows left vocal cord palsy.

**Figure 2:** Chest X-ray postero-anterior view shows increased transverse diameter of cardiac shadow with penciled outline and oligemic lung fields.

**Figure 3:** ECG shows low-voltage and sinus tachycardia.

**Figure 4:** Histopathology slides showing metastatic adenocarcinoma cells in clusters having nuclear atypia.
be interconnected, and their simultaneous presence has added novelty to the case. Presence of drug-resistance made the initial management of the case difficult; however, the organisms were sensitive to another very important first-line anti-tubercular drug INH. Carefully-selected alternative regimen was most probably effective, as evidenced by the negativity of tests for tuberculosis in subsequent follow-up.

Secondary tumours of the pericardium are commoner than the primary ones. The most common secondary malignant tumours are lung cancer, breast cancer, malignant melanoma, lymphomas, and leukemias. Management of malignant pericardial effusion is mostly palliative. Pericardiocentesis may relieve symptoms and establish diagnosis; intrapericardial instillation of sclerosing or cytotoxic agents, baseline systemic antineoplastic therapy and pericardial drainage may prevent recurrence. In the case presented here, systemic anticancer treatment and intrapericardial cisplatin were apparently effective in prevention of frequent recurrence of pericardial effusion. However, he may need pericardial drainage by means of surgical pericardiectomy or percutaneous balloon pericardiectomy followed by further intrapericardial instillation of sclerosing or cytotoxic agents, or immunomodulators in case of future recurrence. Pericardial drainage, when technically possible, is a class-I recommendation in all patients with large malignant effusions because of the high recurrence rate (40 – 70%).

**REFERENCES**