Case Report

Relapsing polychondritis in a patient with Takayasu disease;
A rare association with fatal outcome

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ABSTRACT
We report a case of Takayasu disease that was associated with relapsing polychondritis with an unfortunate outcome. Up to our knowledge, this is the first case to be reported from Jordan with fatal outcome. (Rawal Med J 2010;35:).

Key words: Relapsing polychondritis, takayasu disease, arthrargias.

INTRODUCTION
Takayasu disease (TD) is a chronic progressive occlusive disease of the aorta and its branches, pulmonary artery and renal arteries. A 5-20 year interval may separate acute inflammatory stage and symptomatic arterial occlusive disease. Relapsing polychondritis (RP) is a rare autoimmune disease associated with other autoimmune diseases. The first case of association of RP and TD was reported in 1973. McAdam et al² reported that 25-30% of patients with RP had a concurrent autoimmune disease. They proposed criteria for the diagnosis of RP including recurrent chondritis of auricles, non erosive seronegative polyarthritis, nasal chondritis, ocular inflammation, audio vestibular dysfunction and respiratory tract chondritis.²

The association of RP with a TD have been reported rarely and only a handful cases have been reported so far.³⁻⁴ In 1960, Pearson et al,⁵ introduced the term relapsing polychondritis and later a case of 48-year-old man who had relapsing polychondritis with aortitis mimicking Takayasu's arteritis was reported.⁶ An association of RP and TD was reported in a 7-year-old Lebanese girl was recently.⁷ Aortitis and saddle nose deformity are extremely unusual manifestations of a variety of systemic diseases. A case of saddle nose deformity in a patient with Takayasu's arteritis is presented with rare association of TD and RP with fatal outcome.

CASE PRESENTATION
A 19 year old female presented to the medical clinic in 2003 complaining of fever, fatigue, weight loss 9 kg over 6 months and polyarthritis of hands and feet.

Fig 1. Aortic arch angiogram showing large vessel vasculitis (Takayasu disease).

Her Physical examination at that time showed a Blood Pressure of 120/80 mmHg, Pulse 8/ min regular and Temperature of 37°C. Head and neck examination revealed a pale ill looking lady with right axillary lymph node enlargement. Chest showed vesicular breathing with no added sounds, heart was normal with no murmurs. Abdominal examination showed a soft and lax abdomen and no organomegaly. The peripheral pulses were felt all over.

Fig 2a. saddle nose deformity. Fig 2b. Swelling of the ear auricle.
Her hematocrit was 29% with normochromic normocytic morphology. ESR was 145/1st hour. Serum creatinine, LFT and urine analysis were normal, so were CXR and ECG. Virology screen including hepatitis panel, thyroid function tests, multiple myeloma screen, VDRL test and PPD were negative. Serum iron and folate were normal; ferritin level, however, was high. The rheumatologic screen including immune markers ANA, ANCA, RF were negative, her CRP was 24 (N=0). Abdomen and chest CT scan, upper endoscopy and barium enema were normal. Axillary lymph node biopsy showed reactive changes and bone marrow aspirate showed depleted iron stores with normal cellularity. She was treated as iron deficiency anemia but no improvement and ESR remained 117. In October 2003 she was referred to the Rheumatology clinic with worsening general condition, polyarthralgia, vertigo, difficulty in using her upper limbs, and profound weight loss. Physical examination at this stage showed an ill looking pale young female; there was a systolic murmur over aortic area. CXR, ECG were again normal. 2D Echo, renal ultrasound and brain MRA were normal. Doppler U/S of upper limbs showed stenosis of the aortic arch with bilateral subclavian stenosis. Abdominal and renal angiogram showed vasculitis (Fig 1). She was diagnosed as Takayasu arteritis and started on predinsolone 1mg/kg/daily and methotrexate. One month later, her appetite improved and she gained 5kg weight. Vertigo and claudication had improved. ESR was 19 /1st hour and CRP was negative. She ran a quiescent course for one year and her steroid doses were reduced.

Patient was lost to follow up for six months, when she presented with left sided weakness, tonic clonic convulsions and truncal ataxia. She admitted having recurrent attacks of pain and redness of both ears for the past 6 months, hoarseness of the voice, recurrent eye infections and decreased hearing. Physical examination revealed ill looking thin patient with moon face, bilateral red eyes, and nasal deformity ("saddle nose") (Fig 2a), painful, erythematous, floppy ears with loss of the upper pole of right ear with sparing of lobule (Fig 2b) and absent upper limb pulses. BP in the lower limbs was 130/70 and right sided pyramidal weakness (grade 3/5) was noted. Active synovitis of both hands was found. ENT examination revealed patent upper ways, large septal perforation and ulcerative lesion in upper pole of the auricle.
Her WBC was 10,000, PCV 42, and platelet were 519,000. ESR was 77. Chemistry, LFT and uric acid were normal. ANA 1/360, ANCA, RF, DsDNA were negative. Brain MRI showed periventricular white matter ischemia. Brain MRA was normal; EEG showed intermittent slow sharp wave activity and nasal biopsy showed chondritis. Sinuses CT scan was normal and chest CT scan showed two small nodules. She had right Sensory Neural hearing loss. Ophthalmic examination showed bilateral conjunctivitis.

These new findings were considered as relapse of her disease, with active vasculitis and was started on pulse methyl predinsolone 500 mg for 3 days. Cyclophosphamide infusion, lamotrigine 200 mg 2 BID orally and epanutin 200 mg Q 8 hourly were started. Unfortunately, she developed respiratory decompensation which needed mechanical ventilation. Despite all these measures she succumbed.

**DISCUSSION**

Relapsing polychondritis is a severe, episodic, and progressive inflammatory condition involving cartilaginous structures, predominantly those of the ears, nose, and laryngotraceobronchial tree. Other affected structures may include the eyes, cardiovascular system, peripheral joints, skin, middle and inner ear, and CNS. In this case, our patient presented with vague non specific complaints that took few years before the diagnosis of TD became evident and she developed immune markers. Despite the initial improvement in her clinical and biochemical inflammatory markers, she had a recurrence of her condition that is a natural history of the disease. However, absence of regular follow up and non-compliance made the clinical presentation rather more dramatic. The newer clinical findings of polychondritis and the confirmation of the diagnosis of relapsing polychondritis added to the dramatic outcome.

The association of TD and RP is rare and has been reported only in few cases. \(^4,6-8\) And this case represents another prototype case of this combination that despite no etiological factor is incremented which nevertheless led to grave outcome. In summary, RP is a rare autoimmune disorder was noted to be associated with Takayasu arteritis which is a very rare association. Awareness of the possible manifestations, complications, associated autoimmune diseases and the treatment options will hopefully continue to improve the prognosis and quality of life for patients.
REFERENCES


