PANGERIA

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A case of pangeria, with multiple leg ulcers and premature senility with a history of cataractomy, starting at the age of 6 years, without any history of consanguinity, is reported.

Key Words : Pangeria, Aortic calcification

Introduction

Pangeria, also known as Werner's syndrome, is a rare inherited disorder in which the ageing process is accelerated. It is characterised by short stature, senile appearance, cataracts, joint contractures, early menopause, various skin changes (including premature canities, baldness and ulceration) and an increased risk of malignancy.[1] This syndrome is due to an autosomal recessive gene, with a calculated gene frequency of 1 to 5 per 1000 population.[2]

Case Report

A 25-year-old Hindu male patient presented with multiple ulcers on the left foot. Patient's general appearence showed marked senile changes (Fig.1). He was only 110 cm tall and weighed 17 kg. He developed greying of hairs at the age of 6 years. He also had cataract removal of both the eyes at the age of 7 years. His face showed wrinkling, beaked nose, circumoral radial furrows, taut lips and he had a peculiar hoarse voice. Patient had a general loss of muscles and subcutaneous fat. Extremities were slender with hide-bound skin and hands and feet were smaller in size. Genitalia were normal except size of the testes and penis were smaller for the age. Patient had right elbow contracture, other joints were normal. Skin in general showed focal hyperpigmentation. Nails were rough and mild degree clubbing was evident. Axillary hair, beard and moustache were absent and pubic hairs were sparse. Patient had normal intelligence and no sign of any apparent malignancy any where in the body. There was no history of consanguinity in the family.

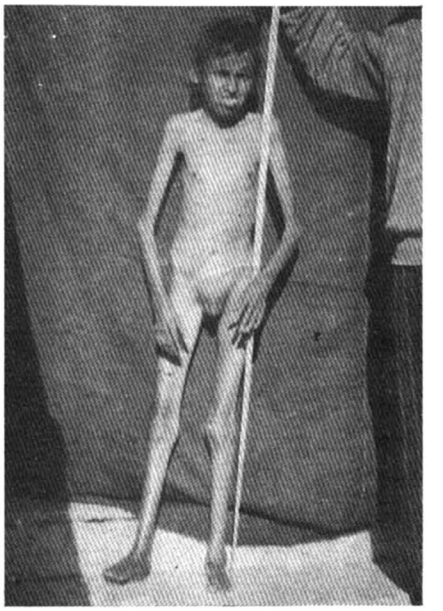

Fig. 1. Pangeria.

Laboratory investigations showed otherwise normal haemogram except an ESR of 38mm/hour. Blood sugar (random) was

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normal, serum calcium was 13.2 mg%, serum cholesterol 215 mg%, serum alkaline phosphatase 21 KAU. Skeletal survey revealed generalised osteoporosis with calcification of multiple ligaments and calcification of the arch of aorta and descending aorta (Fig. 2). Doppler study showed absence of both right and left arteria dorsalis pedis pulsation, pulsation in other arteries was normal. ECG showed small complexes in leads II, III, avF and P-pulmonale in lead II. Ultrasonography of the abdomen revealed no abnormality.

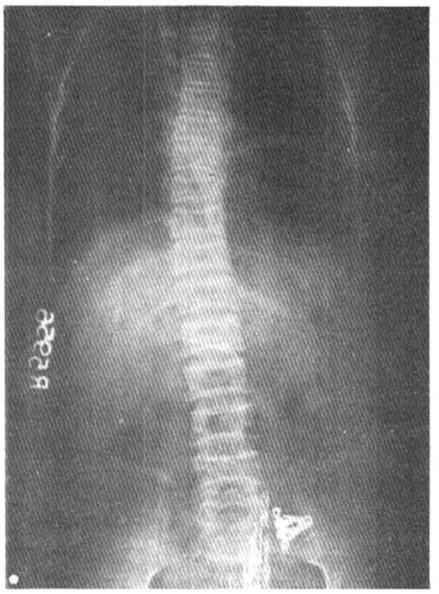

Fig. 2. Chest radiograph showing calcification of the arch of aorta and descending aorta.

Discussion

Pangeria is a rare premature ageing syndrome and about 200 cases have been described in the world literature till 1989. Parental consanguinity and occurrence of this condition in the siblings have been reported.[3] The condition usually starts at the age of 14-18 years, but may rarely be present as early as 8 years of age.[2] The patient in the present report showed almost all the classical changes of pangeria and had calcification of the aorta. The spontaneous occurrence, early age of onset and calcification of the aorta remain the interesting features of this case.

References

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