

PSEUDOXANTHOMA ELASTICUM (A CASE REPORT)

By

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Pseudoxanthoma elasticum is a rare heritable systemic disease of connective tissue (elastic tissue) mainly affecting the skin, eyes and the blood vessels. It first attracted attention as a skin disorder in 1884 when Balzar in France first described the clinical and histological changes in skin in an autopsy case of a tuberculous patient. Darier in 1896 defined the disease as a separate entity and suggested the name PXE. The disorder is an uncommon one as indicated by the fact that prior to 1940 only 125 cases of skin and ocular changes and 68 cases of skin changes alone were described (Mc Kusick, 1960). and there have been less than 400 cases reported in the world literature since its first description (Karl & Siefert, 1964). Only a few cases have been reported in India (Thambiah et al, 1964; Krishnan Kutty and Sadasivan, 1965; Muley and Bikhchandani, 1965; Velou et al, 1966 etc.) A case seen in Srinagar Medical College recently is reported below :

CASE REPORT

K. D. 49 year male was admitted as a case of Jaundice for past 15 days. He complained of pain in right hypochondrium with intermittent fever and rapid deterioration of health. He made no mention of any skin lesions. He was married, had two normal children and there was no history of consanguinity of marriage in parents. There was no history suggestive of PXE in the family.

The patient looked ill and slightly emaciated. He had jaundice and pitting oedema on right lower part of chest. Characteristic skin lesions were present on the sides and front of neck and in periumbilical region in the form of small soft yellowish coloured papules which at places had coalesced together into plaques and these were more or less parallel to the folds of neck, had a reticulate pattern around the umbilicus. No such lesions were detected elsewhere on skin or mucous membranes. The lesions did not show any induration. There were also some purpuric lesions on the thighs and buttocks.

Systemic Examination :- Liver was 3 fingers below the right costal margin and tender with upper limit of dullness in the right 3rd intercostal space. No other abnormality was detected on systemic examination. B. P. was normal. All peripheral pulsations were normal.

Fundoscopy : No abnormality detected.

INVESTIGATIONS

Blood : Hb. 6 gms%, TLC 12600/cmm, polys. 73%, Lymphos. 23% Eosinos 3%, Monos1%. Urine : Urobilinogen present, Bile pigments present. Stools : Ova

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of *Ascaris lumbricoides* present. Prothrombin times: 22 seconds (control 20 seconds). Liver function tests: Total proteins 6gms%. Alb. 3gms%. Glob. 2gms%, Serum alkaline phosphates 17KA units Serum bilirubin 4.6mg, thymol turbidity 4 units, SGOT: 100 units per ml. SGPT: 80 units per ml. Fluoroscopy: Right dome of diaphragm was much elevated and its movements restricted. Gas present under the dome of diaphragm.

X-Ray: Confirmed the presence of gas with fluid level under the right dome of diaphragm. E. C. G : Normal.

The patient was operated, subphrenic abscess drained and a round worm was taken out of the common bile duct. There was no evidence of generalized peritonitis. The patient however died on fourth post-operative day.

Biopsy of skin lesion : Pseudoxanthoma elasticum.

DISCUSSION

PXE which is a degenerative disorder of elastic tissue of body may commence at any age though it develops most frequently between second and fifth decade of life (Whitcomb and Brown, 1962). Skin alone may be involved and this occurs in about 10% cases (Beeson and Mc-dermott, 1964). Such patients are usually detected during examination for some other disease or due to the presence of disease in other family member, such cases besides present patient are reported by Whitcomb and Brown (1962), Muley and Bikhchandani (1965). This is usually because of the asymptomatic character of the skin lesions. Cardiovascular abnormalities have been reported in about 80% cases and skin with eye changes in 60% cases (Beeson and McDermott, 1964) Sandbacka and Holmstrom (1939) in a review of 100 cases reported angioid streaks in 87% cases. While Whitcomb and Brown (1962) reported angioid streaks in 8 of 12 patients. Angioid streaks are believed to be more common in males while skin lesions more in females (Beeson and McDermott, 1964). However no eye lesions were detected in present case. History of consanguinity in parents which was absent in present patient is reported by Velou et al (1956) and Muley and Bikhchandani (1965) in one of four cases. Similarly familial prevalence of the disease which was absent in present case was reported by Whitcomb and Brown (1962), Muley and Bikhchandani (1965) and Velou et al (1966).

SUMMARY

A case of Pseudoxanthoma elasticum with characteristic skin and purpuric lesions detected during the course of investigations for Jaundice with no other cardiovascular and ocular manifestations, no family history of PXE and no history of consanguinity of marriage in parents, is reported. Literature is briefly reviewed.

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BIBLIOGRAPHY

1. Balzer, F., Recherches sur les caracteres anatomiques du xanthelasma. Arch. dephysiol., 4:65, 1984 (Quoted by 9).
2. Beeson, P. W. and Mc Dermott, W., Pseudoxanthoma elasticum. Cecil-Loeb, Text book of Medicine 11th Ed. W. B. Saunders Co. Philadelphia and London, 1964. pp 520.
3. Connor, P. J., Juergens, J L., Perry, H. O., Hollenhorst, R. W and Edwards J. E., Pseudoxanthoma elasticum and Angioid streaks. Am. J. Med., 30:537, 1961.
4. Darier, J., Pseudoxanthoma elasticum. Mschr. Prakt. Derm., 23:609, 1896 (Quoted by 13).
5. Krishnan Kutty, P. K. and Sadasivam; P. B., Pseudoxanthoma elasticum. J. Ind. Med. Ass. 44:200, 1966.
6. Karl, C. and Siefert, F. E., Pseudoxanthoma elasticum with sickle cell anaemia. Arch. Int. Med., 113:135, 1964.
7. Mc Kusick, V. A. Heritable disorders of connective tissue 11th. Ed. C. V. Mosby Co. St. Louis, 1960 pp. 213 (Quoted by 3).
8. Muley, D N. and Bikhchandhani, R., Pseudoxanthoma elasticum (4 case reports). Ind. J. Derm. Vener. 31:75, 1965.
9. Robertson, M. G. and Schroder, J. P., Pseudoxanthoma elasticum A systemic disorder. Am. J. Med., 27:433, 1959.
10. Sandbacka-Holstrom, Das Gronbiad-strandbur' sche syndrome. Pseudoxanthoma elasticum Angioid streaks. Acta. Dermatovener., 20:684, 1939.
11. Thambia, A. S. Rao. U. S. and Annamalai, R. J. Ind. Med. Profession, 10:4783, 1964 (Quoted by 12).
12. Velou, A. Sardarilal, Mathai, G. and Balasubramanyam, M., Pseudoxanthoma elasticum (report of 3 cases). Ind. J. Derm. Vener. 32:182, 1966.
13. Whitcomb, F. F. and Brown C. H. Pseudoxanthoma elasticum. (case reports of 12 patients). Ann. Int. Med. 56:834, 1962.

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