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Malignant Haemangiopericytoma

A case report

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HAEMANGIOPERICYTOMA is a rare vascular tumour of universal distribution. Diagnosis is mainly by histopathological evidence. The rarity of reports of this condition in Indian literature makes us enthusiastic to publish this case note.

Case Report

U.N., a male aged 85 years, was admitted in the Medical College Hospital, Calicut on 9-9-1967 with a soft, dark blue, haemorrhagic swelling in left axilla for one month. Physical examination revealed no other abnormality, except grade I benign enlargement of prostate. To control bleeding, the tumour was excised and sent for biopsy report. Patient was discharged at request on 13-9-67, but was re-admitted on 27-9-67, with similar haemorrhagic swellings in right axilla and left supraclavicular region. They were removed on 29-9-67 and sent for biopsy. They had no capsules. By this time there were multiple swellings in both axillae, intercostal spaces, left deltoid region, supraclavicular fossae, left inguinal region, both sides of neck, anterior abdomnal wall, scapular region, right buttock and paravertebral region. Swellings were of variable size, ranging from 2 cm. to 12 cm. in diameter. (Fig. 1). While recent ones were subcutaneous, firm and mobile, older ones were cystic, fixed and with a tendency to infiltrate the skin. All were non tender. During his stay in hospital some swellings were rapidly growing, while others were stationary. New ones were also appearing. On 16-10 67, hoarseness of voice was noticed. Laryngoscopy revealed recurrent nerve paralysis of right side, which could be explained by pressure from cervical swellings. On 25-10-67, a fungating swelling in right suprascapular area was removed and sent for histological study. On 28-10-67, right sided supranuclear type of facial paralysis was detected without any other nerve involvement, which became bilateral towards 4-11-67. From 7-11-67 onwards, he was mentally depressed and confused. Next day weakness of both lower limbs with an equivocal plantar response was noticed. By 9-11-67 he was incontinent for urine and faeces. On 10-11-67 he was discharged on request. At discharge, there were more than 25 tumours on his body. He was semiconscious, unable to walk or talk. No follow up could be made later on. During his stay in hospital Endoxan (Cyclophosphamide) 200 mg daily I.V. was given for 10 days, emperically. No beneficial results were obtained.

Investigations

Urine : Alb : nil ; Sugar : nil ; Microscopy : RBCS plus ; Pus cells : nil ; Granular casts : nil.

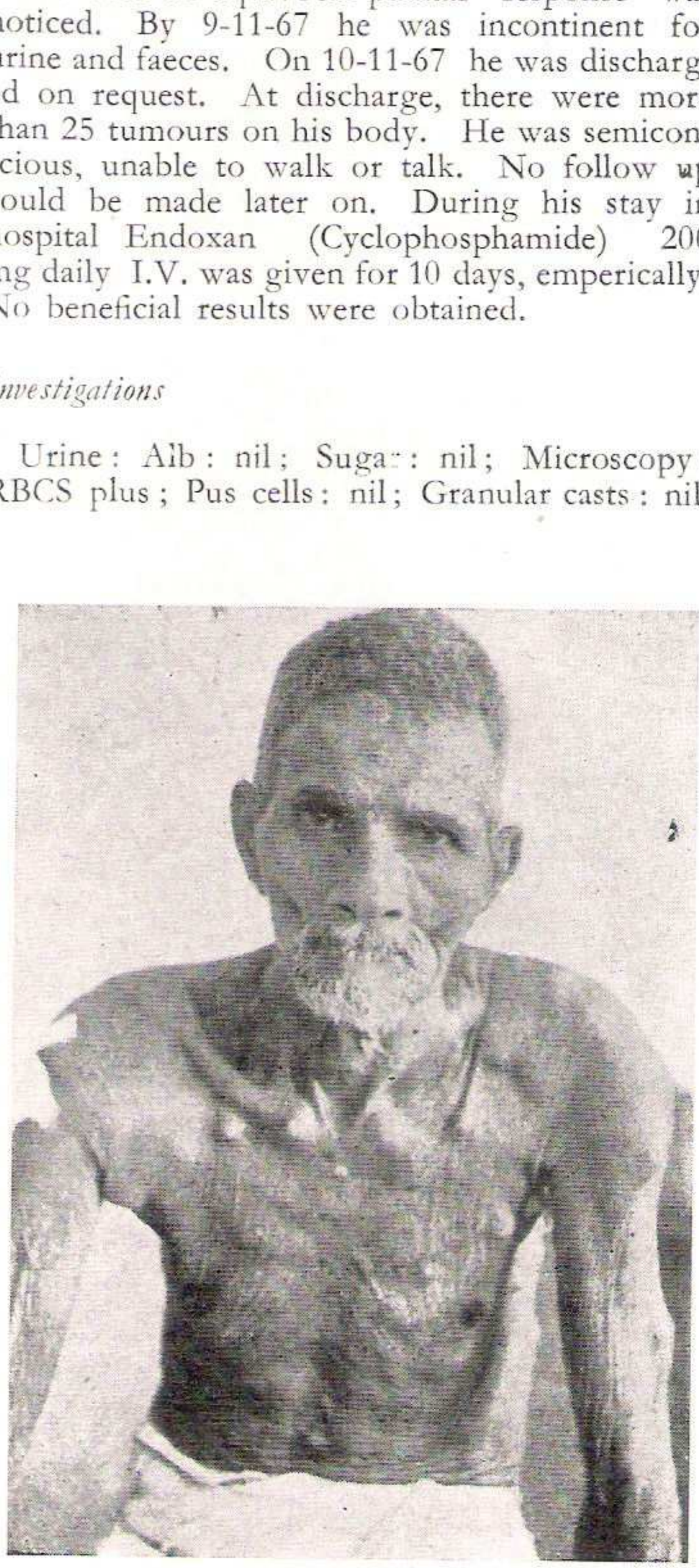

Fig. 1

Patient : Note swellings of different sizes.

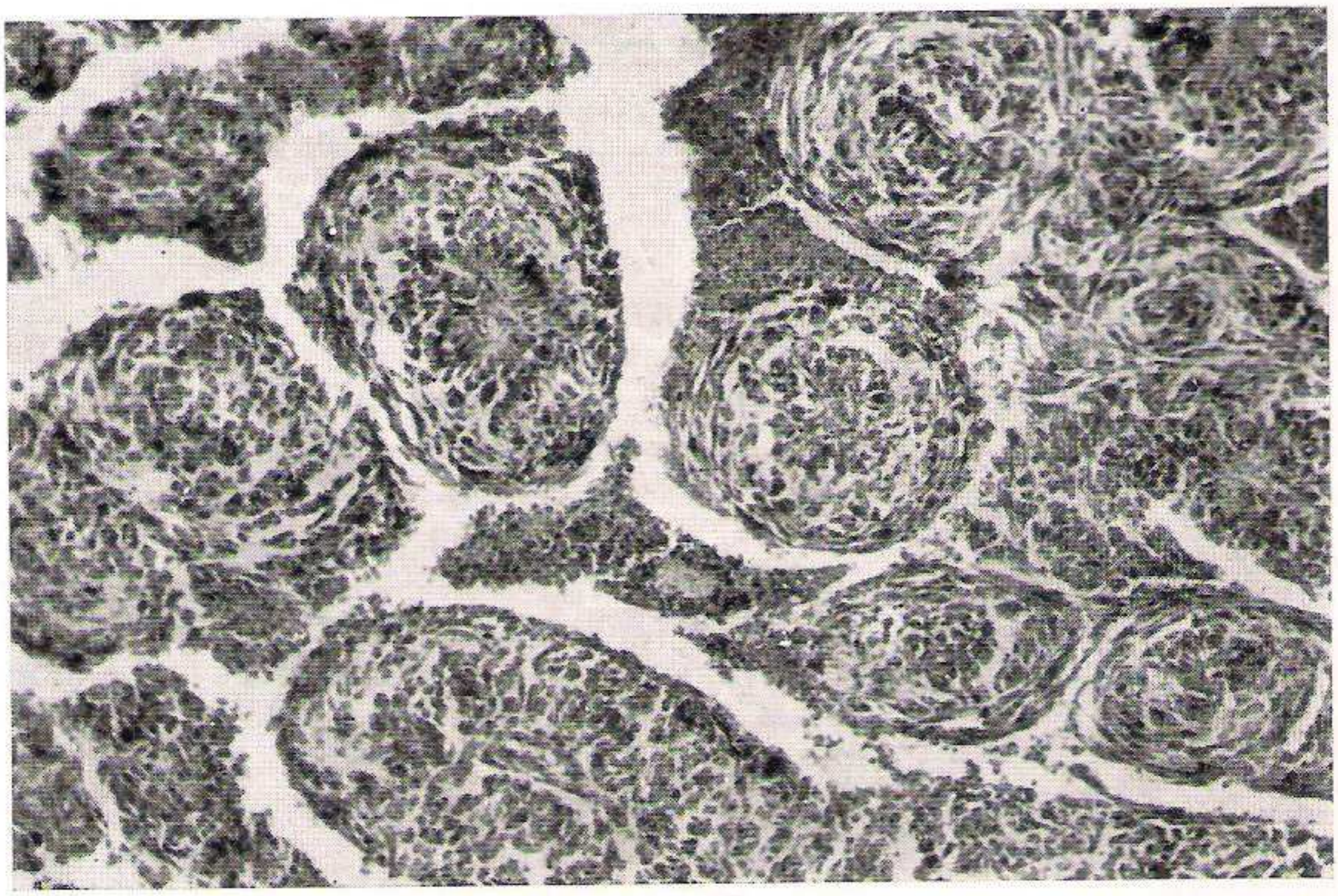

Fig. 2

Haemangiopericytoma H & E Staining Low power.

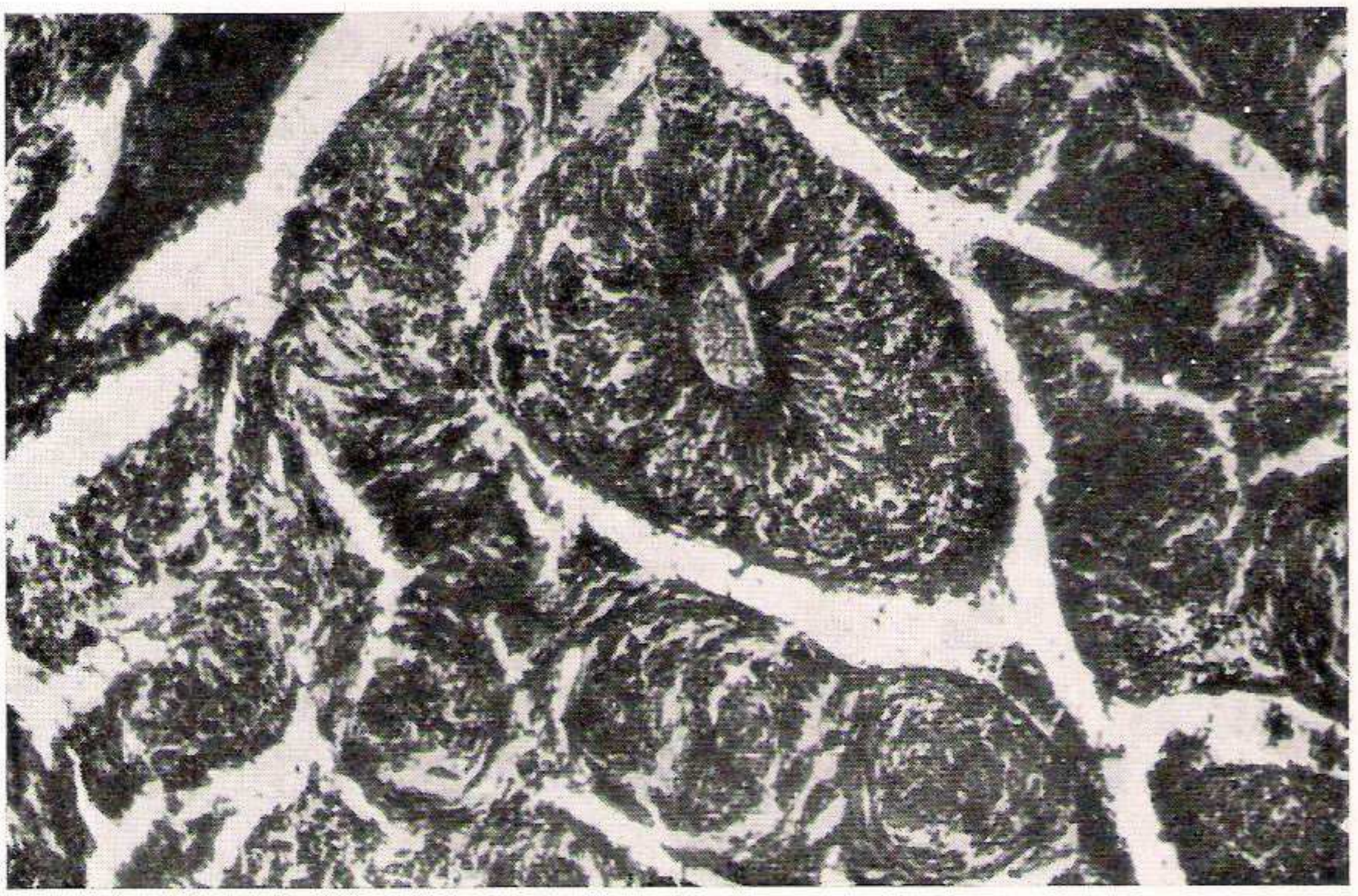

Fig. 3

Haemangiopericytoma Retieulin Stain Low power.

Blood : TC : 6,550/cu. mm. DC : P_{69}, L_{22}, E_q, No abnormal cells. ESR : 40 mm/h. Total proteins : 7.3 G %; Alb : 4.3 G %; Glob : 3 G %. Blood urea : 25 mg %.

X-ray chest : No definite intra-thoracicl esion. I.V.P. Report : (9497/10-11-67) : Multiple nodules in the kidney disfiguring the calyceal pattern, especially on left side.

Biopsy Reports : Nos. 2257/67 and 2562/67. Macroscopy : Cut section shows dark brown areas. Microscopy : (H & E) (Fig. 2): Tumour tissue contains large number of capillary spaces, some of which contain RBCs. Tumour cells are polygonal with large hyperchromatic vesicular nucleus and are seen arranged radiating from the endothelial lining of capillary spaces. Demarcation between different clumps of cells is present. There are areas where the tumour cells are spindle shaped and are arranged in sheets. Some areas show haemorrhage and macrophages carrying haemosiderin pigment. Tumour tissue is seen infiltrating the surrounding tissues and there is no definite delimitation. Reticulin stain (Fig. 3) shows the perivascular arrangement of tumour cells. Reticulin sheaths are intact. Report : Haemangiopericytoma.

Discussion

The pericyte was originally described and named by the Swiss histologist, Zimmermann in 1923. Haemangiopericytoma was first described by Stout and Murray in 1942. The tumour was reported to be arising from superficial soft tissues (McCormack and Gallivan, 1954; Wise, 1952; Forrester and Houston, 1951), meninges (Begg and Garret, 1954), orbit (Goodman, 1955; Fox, 1955), thoracic cavity (Fergeson *et al*, 1954), ileum (Smith and Swenson, 1954), Sigmoid (Ault *et al*, 1951), rectum (Kay and Warthen, 1953) and uterus (Pedowitz *et al*, 1954; Tupper, 1957). Kauffman and Stout (1960) presented 31 cases of haemangiopericytoma occurring in children below the age of 15. Radiological aspects were discussed by various authors (Kent, 1957; Mujahed *et al*, 1959; Friedman and Egan, 1960; Fink and Oberman, 1963; Feldman and Seaman, 1964). Recent reports include 5 cases by Vidrine and Welsh (1964) and 24 cases by O'Brien and Brasfield (1965). Cases of haemangiopericytoma associated with hypoglycemia were reported by Crocker and Veith (1965) an Paullada *et al* (1968) with review of literature. Lee and Kay (1962) described one renal haemangiopericytoma with reviews of 2 such cases. Indian literature consists of one case report by David and Laxminarayanan (1959) and 2 cases by Tandon *et al* (1964). Haemangiopericytoma is demonstrated in dogs also (Innes, 1958).

Clinical features

No age or sex prediliction is noticed. It is reported from new born (Kauffman and Stout, 1960) to 92 year old (Stout and Cassel, 1943, cited by Kent, 1957). Peak incidence is in 4th decade of life (McCormack and Gallivan, 1954; O'Brien and Brasfield, 1965). Theoritically the tumour can occur wherever capillaries exist. Usually it presents as a single painless subcutaneous swelling of a few centimetre in diameter; but cases are recorded in which tumours occupying half the pelvis of pleural space (Feldman and Seaman, 1964). Tumours may remain stationary, or may grow very slowly; while some are rapidly growing. Some show local invasiveness and local recurrence; while others are presented with distant metastases. In the majority of cases, duration is months to a few years. Stout and Cassel (*loc. cit*) reported an omental tumour present for 60 years. Vascularity is inconspicous in some cases; while others show marked vascularity. Cut section is dark brown, yellow or tan in colour. Some tumours have true capsules, some have pseudocapsules, while others have no capsules. Necrosis is seldom reported. Calcification of soft tissue tumours may occur (Kent, 1957; Mujahed *et al*, 1959).

Pathology

Histologically the basic pattern is the proliferation of capillaries lined by flattened endothelium with an intact but proliferated reticulin sheath, surrounded by the pericytes placed on a supporting meshwork of reticulin fibres. Capillaries are usually prominent; if not, they can be rendered conspicuous by reticulin staining, which also demonstrates the perivascular orientation of tumour cells. According to McCormack and Gallivan (1954), most immature tumours comprise of uniform small cells with sparse cytoplasm and oval nucleus with frequent mitotic figures; while most mature forms are composed of spindle cells with abundant oxyphilic cytoplasm with a few mitotic figures.

Two closely allied conditions to haemangiopericytoma are haemangio-endothelioma and glomus tumour. In haemangio-endothelioma tumour cells are found inside the capillary sheath, whereas in haemangiopericytoma cells are characteristically outside the sheath. In glomus

tumour, groups of cells are enclosed by reticulin fibres; while in haemangiopericytoma, tumour cells completely fill the space between capillaries without any interruption, thus lacking the neuromyoarterial component of the glomus tumour.

It is possible that pericyte is stimulated by hypertension (McCormack and Gallivan, 1954). Many authors (Forrester and Houston, 1951) do believe that the pericyte is a modified smooth muscle cell; but others (McCormack and Gallivan, 1954) are of opinion that it is neither fibrocytic nor myocytic in derivation. Proliferation of vessels is not neoplastic, but compensatory; an attempt to supply the nutritional requirements of an increasing mass of tissue (Begg and Garret, 1954).

Biochemical analysis (McCormack *et al*, 1961, cited by Feldman and Seaman, 1964) revealed high concentration of acetyl-choline in these tumours. Pericytes may be sensitive to acetyl-choline and since vessels are surrounded by pericytes, the tumour is subject to vascular dilatation or canalisation of its potential vascular spaces. Biochemical and electrone microscopic studies revealed overstorage of glycogen in one case of haemangiopericytoma (Paullada *et al*, 1968).

Age of the patient or encapsulation of the tumour cannot be taken as a criterion for malignancy. No congenital tumour is known to have been malignant (Kauffman and Stout, 1960). Haemangiopericytoma of thigh and calf are very malignant; while those of mouth are of low graded malignancy (Vidrine and Welsh, 1964). Histological diagnosis of malignancy is difficult; although an unusual number of mitoses, vesicular or vacuolated appearance of nuclei, disorderly pattern of reticulin fibres and comparative decrease in number of vessels are all in favour of malignancy (Tandon *et al*, 1964). Most important deciding factors are the presence of local invasion, local recurrence and distant metastasis. Metastasis may occur by way of blood or lymph streams. Longest interval between removal of a primary thoracic tumour and recurrence was 26 years (Feldman and Seaman, 1964). The tumour is highly malignant, not in a usual 5-year concept; but over a life time (O'Brien and Brasfield, 1965). Malignancy is 1 in 5 according to Lee and Kay (1962); Kauffman and Stout (1960) reported malignancy as 35.5 per cent; it is 56.5 per cent in the series of O'Brien and Brasfield (1965); while according to McCormack and Gallivan (1954), it is 50-70 per cent.

Treatment

Wide excision is the treatment of choice. Regular long-time follow-up is essential in view of the frequency of delayed recurrence. Recurrence is treated by radical excision plus irradiation. Partial or complete regression of metastases may be achieved by irradiation. These tumours are generally radioresistant; lethal tumour dose is 7,500 to 9,000 rads in 30-60 days (Friedman and Egan, 1960). Actinomycin D, Cyclophosphamide and Mitomycin C gave no objective response to cases of O'Brien and Brasfield (1965); whereas promising results were attained with Nitrogen Mustard by Fink and Oberman (1963).

Comments

A unique feature in the clinical presentation of our case is the appearance of multiple subcutaneous nodules within a very short time. In this case a particular primary site cannot be diagnosed, for all the nodules appeared almost simultaneously. Rapid deterioration of the general condition of the patient is also remarkable. Mental and motor involvement suggest a possible brain metastasis, which should be extensive in nature. I.V.P. is suggestive of renal involvement. It is worth noting

that reported cases of renal haemangiopericytoma are very few in literature. Vascularity was conspicuous in these lesions. In our case Cyclophosphamide was virtually useless.

Summary

1. A case of malignant haemangiopericytoma is presented, main feature being multiple subcutaneous swellings.

2. A brief review of available literature is given.

3. Clinical features, pathology and treatment are discussed in general terms.

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