Wermer Syndrome

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Case Report

M.I. a 35 years old man was referred to us with polyuria, polydepsia, severe backache, difficulty in walking, vomiting and a swelling in the left upper molar region. Previously he was operated for left sided renal stone at Saidu Hospital, Swat in 1985. Then he developed a swelling in the left lower molar region in 1986. Biopsy revealed Giant Cell Reparative Granuloma. A year later he developed a hard painful lesion near the upper end of left tibia. Biopsy showed Giant Cell Lesion. In March 1990 he developed another swelling in the left upper molar region; biopsy again revealed Giant Cell Reparative Granuloma. He also has a positive history of Lipomas in the family.

Examination

He looked ill, dehydrated with multiple cutaneous lipomas. He had generalized weakness of all muscle groups especially the proximal muscles of the lower limbs. He was investigated for Hyperparathyroidism and was found to have biochemical abnormalities, including high serum PTH levels. Thyroid scan showed a hot nodule in the lower pole of the right lobe. Parathyroid subtraction scan (Thallium 201) confirmed a parathyroid adenoma behind the upper pole of the right lobe of thyroid gland. He went through

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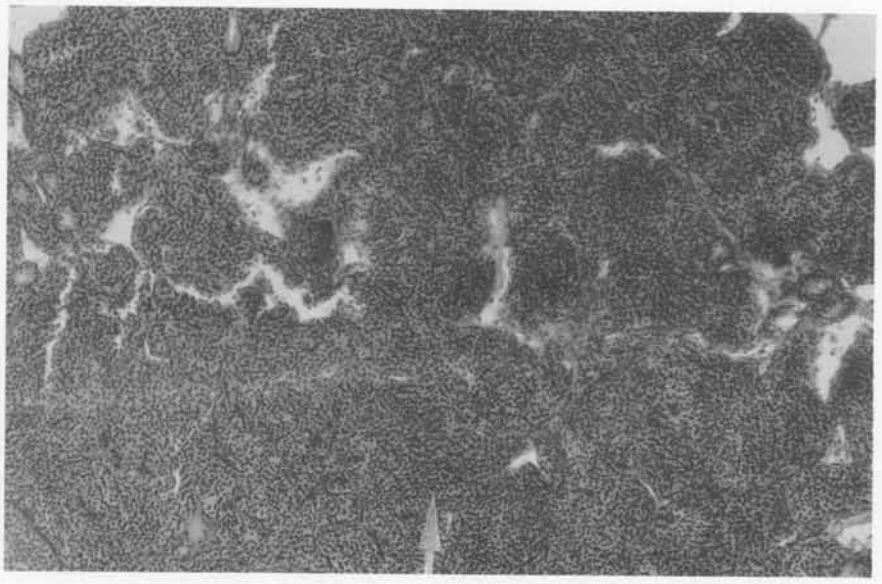
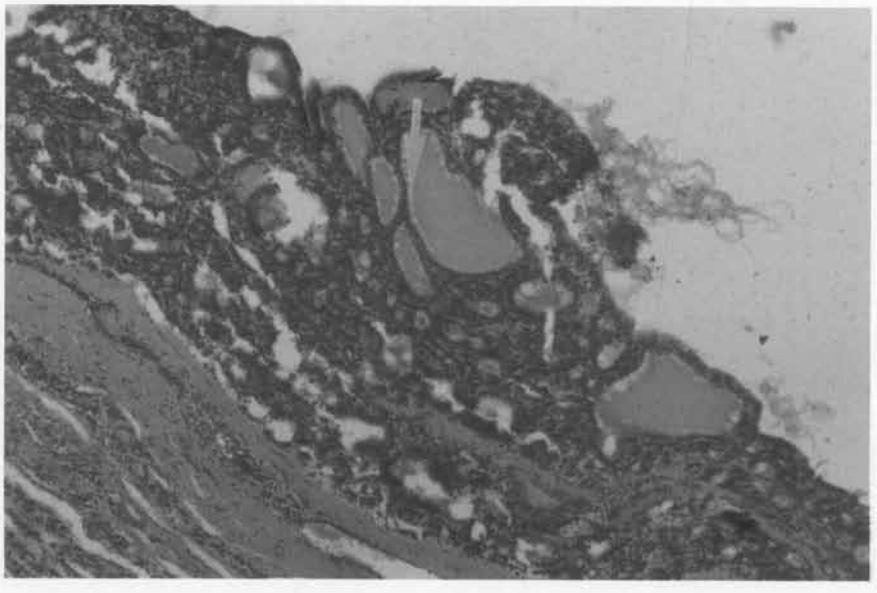
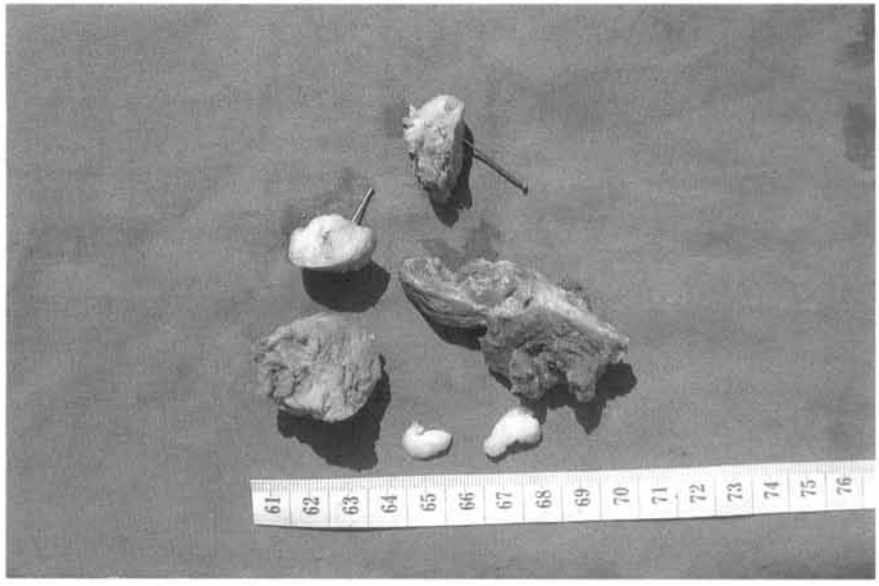


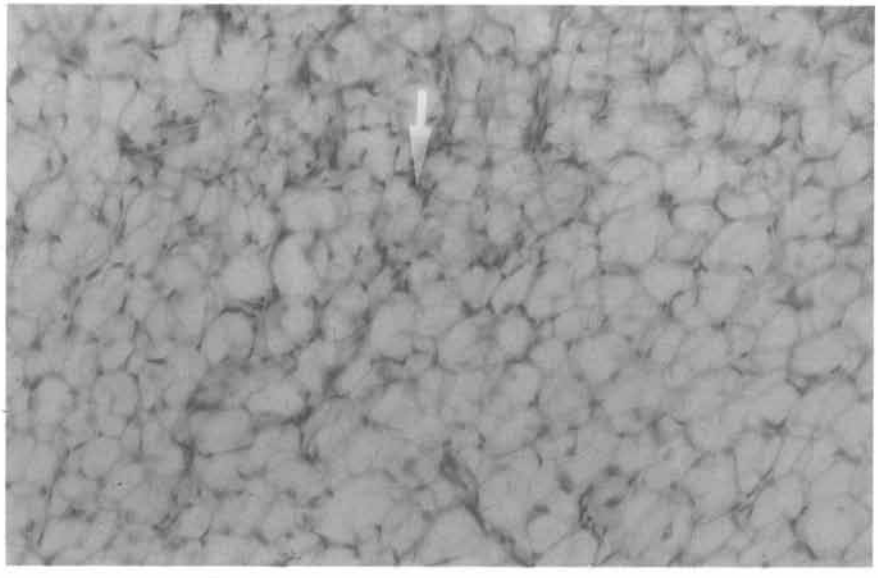
1. PARATHYROID ADENOMA, THYROID ADENOMA, LIPOMA
Giant Cell Tumour



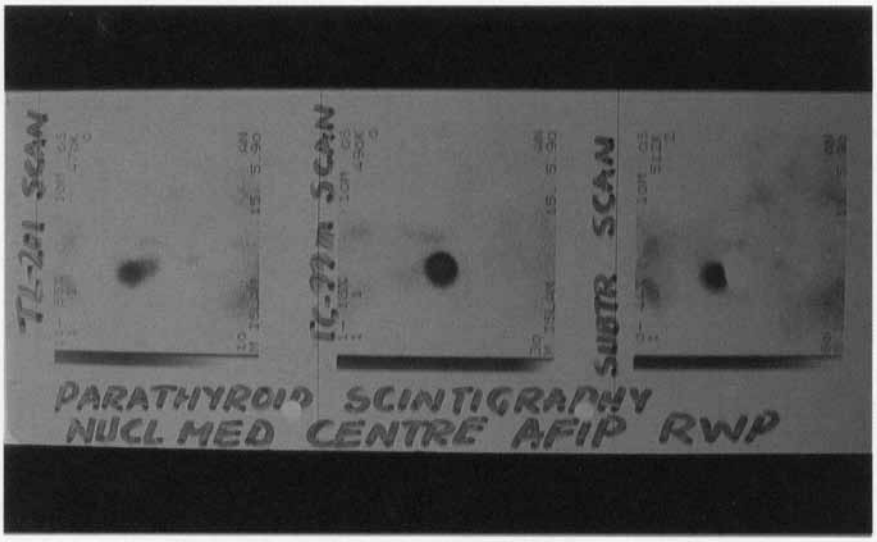
2. THYRIOD NODULE HISTOLOGY



3. PARATHYROID NODULE HISTOLOGY



4. LIPOMA HISTOLOGY



5. THALIUM-201 SUBTRACTION SCAN

surgery. Operative findings confirmed parathyroid adenoma, thyroid nodule, and lipomas. Histopathology of these lesions also confirmed these findings.

The combination of parathyroid adenoma, thyroid nodule and multiple lipomas constitute WERMER SYNDROME (MEN-1).

A follow-up study of one year showed complete recovery with no clinical or biochemical abnormalities.

Discussion

Wermer syndrome (Multiple Endocrine Neoplasia Type I) was first described by Wermer in 1954. It is inherited as an autosomal dominant triat.[1] Glandular involvement is as follows:

Parathyroid gland: 90-95%
Pancreatic islet cells: 30-36%
Adrenal cortex, thyroid: rare[2].

Fifty percent patients have 2 or less than 2 glands involvement while 20% have 3 or more than three. Involvement may be simultaneously or successively. Etiology of this syndrome is not known exactly though some have suggested neural crest cell abnormalities and recently there is a new evidence for a circulating growth factor as the basis for the syndrome[3].

Glandular Involvement

Parathyroid involvement is an essential component of the syndrome[1]. Usually this involvement is silent for years and in most of the cases all glands are involved. Pancreatic islet cells abnormality may present with any hormone secreting adenoma. Most commonly gastric acid secretion with Zollinger-Ellison syndrome is seen[4]. Adenomas or hyperplasia of pituitary with GH or prolactin secretions are seen in 20% cases. Rarely adrenal cortical or thyroid adenomas are found.

Presentation

Most of the patients present beyond 60 years of age. Hypercalcaemia, kidney stones and peptic ulcer are the commonest manifestations. Hypoglycemia, acromegaly, hyperprolactinaemia, galactorrhoea, amenorhoea and adrenal or thyroid hyperfunction may be seen. Rarely patient presents with lipomas, hepatomegaly or carcinoid syndrome[5]. Association with schwannomas, thymomas, carcinoid tumours, inclusion cysts or cutaneous leiomyomas have been reported[6]. Once diagnosed, it needs family screening of the patient as well as follow-up to detect any glandular involvement that arise. Treatment is according to the glandular involvement.

References

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