Ingestion of shark liver associated with pseudotumour cerebri due to acute hypervitaminosis A

Sir: The syndrome of benign intracranial hypertension or pseudotumour cerebri resulting from hypervitaminosis A was first described by Gerber et al in 1954.1 Acute hypervitaminosis A following the ingestion of polar bear liver is known to occur and arctic explorers have manifested the symptoms of pseudotumour cerebri viz. severe headache, vomiting, drowsiness and irritability a few hours after the ingestion of polar bear liver, as reported by Elisha Kane in an anecdotal account of arctic exploration in the 19th century.2

Pseudotumour cerebri due to acute hypervitaminosis A associated with the ingestion of shark liver (Scoliodon sp.) has not been previously documented although Lonie in 1950 described violent frontal headache, nausea, vomiting, vertigo, drowsiness and irritability in adults following the ingestion of shark liver.3 Indeed a recent review article on pseudotumour cerebri could not identify a single adequately documented case where acute hypervitaminosis A was the aetiological factor.4 We therefore report a case of pseudotumour cerebri due to acute hypervitaminosis A following the ingestion of shark liver.

A 25-year-old previously well housewife was transferred from a peripheral coastal hospital to our department with a history of persistent headache, vomiting and diplopia developing over a period of one week following ingestion of a meal of cooked shark liver. All the other family members too who had partaken of this meal had developed those symptoms thus necessitating admission to the local hospital but had recovered with symptomatic treatment within a few days. The persistent nature of our patient's symptoms coupled with the discovery of bilateral papilloedema resulted in her referral to the Neurology Unit for exclusion of a possible intracranial space-occupying lesion. Examination revealed an afebrile, healthy looking, slim, alert female with bilateral florid papilloedema, enlarged blind spots and bilateral partial abducens palsies. There were no other abnormalities. Plain skull radiographs, both posteroanterior and lateral views, were normal there being no evidence of longstanding raised intracranial pressure; the EEG, bilateral carotid angiography and myodil ventriculography too did not reveal any abnormality (facilities for computed tomography are presently unavailable in Sri Lanka).

Analysis of the ventricular cerebrospinal fluid obtained at the time of ventriculography revealed no abnormality. Routine haematology, liver function tests, urine analysis and serum electrolyte estimation including serum calcium were within normal limits. Estimation of serum vitamin A levels using the spectrophotometric method of Neeld and Pearson revealed markedly elevated levels of vitamin A: 177.3 g/dl (normal range 35–70 g/dl) while the β-carotene levels were within normal limits: 87.46 g/dl (normal 50–200 g/dl).5 Hepatic vitamin A levels were determined by the fluorometric method of Thompson et al on a liver biopsy specimen obtained using the Menghini needle. The hepatic vitamin A content of the patient was 18 mg/100 g liver tissue (fresh weight) while the mean value for healthy adults in Sri Lanka was 10–77 ± 1–2 mg/100 g liver tissue (TMS Atukorala, M JN, Thamotheram, unpublished observations). The patient made an extremely satisfactory recovery over a period of eight weeks and at the time of discharge was asymptomatic; there was no external ophthalmoplegia, blind spots were normal in size and the papilloedema had receded while an EEG revealed no focal or paroxysmal activity. Serum vitamin A levels estimated prior to discharge was within normal limits: 68 g/dl. The liver biopsy, was not repeated prior to discharge on account of the patient's excellent recovery.

The shark species Scoliodon abounds in the coastal waters around the island of Sri Lanka and is frequently consumed by coastal dwellers. The vitamin A content of fish liver in general varies between 2000 and 100,000 IU vitamin A/g, with the shark liver in particular containing one of the highest concentrations of vitamin A.6

Our patient who consumed approximately 150 g (5 oz) of shark liver would therefore have ingested at least 7,650,000 IU of Vitamin A. It has been estimated that daily ingestion of 100,000 IU or more of vitamin A over a period of a few months is required to produce the pseudotumour cerebri syndrome.7 Our experience would support this view, although information about this patient's previous vitamin A status was not available. Further supportive evidence of the fact that the origin of this patient's markedly elevated serum vitamin A levels was from an animal source was shown by the fact that her serum β-carotene levels were not elevated, thus virtually excluding the possibility of the vitamin A being derived from a plant source rich in β-carotene, eg. carrots, mangoes, papaw.

While the mechanism of papilloedema in hypervitaminosis A remains hypothetical, it must be emphasised that it is indistinguishable from the papilloedema due to a brain tumour. Pseudotumour cerebri, therefore remains a diagnosis of exclusion. The above case highlights the need for the physician to be alert to the possibility of acute hypervitaminosis A induced pseudotumour cerebri in areas where shark liver is consumed. Furthermore in cases of pseudotumour cerebri where no obvious aetiological factor can be identified, inquiry into the patients' dietary history may prove worthwhile.

References


2 Kane EK. Arctic explorations in the years 1853, 1854, 1855. Philadelphia: Childs & Peterson Pub. 1856; Vol 1: 392.


Thymolipoma in association with late-onset myasthenia gravis.

Sir: The value of steroid treatment and thymectomy in patients with myasthenia gravis without thymomas, especially in
late-onset cases, has yet to be clarified.\(^1\)\(^2\) Olanow et al.\(^3\) reported excellent results with thytenomy in 12 patients with late-onset myasthenia gravis. One of these had a thymolipoma. This tumour has, to our knowledge, only been reported in the world literature as a cause for myasthenia gravis in two other cases.\(^4\)\(^5\) We therefore present the fourth case in this paper.

In May 1980 a 61-year-old man noted diplopia for the first time. In September he complained of weakness of the jaw and neck muscles, dysarthria and problems with swallowing. The muscles of the limbs were not affected, but there was weakness of the sphincter ani muscles with incontinence. The diagnosis was substantiated by observing the response to edrophonium chloride. Chest radiographs showed no mediastinal abnormalities. Thytenomy was discussed, but not performed. Treatment with pyridostigmine 250 mg per day improved the myasthenic symptoms apart from the weakness of the sphincter muscles. In July 1981 the patient made a spontaneous recovery and for three months no medication was needed. Then he relapsed, at which time the limbs also were affected. In October 1981 treatment with dexamethasone 16 mg per day was given for 10 days, and until January 1982 he did well on pyridostigmine 200 mg per day. Thereafter his myasthenic symptoms worsened, and treatment with prednisone was initiated in an intensive care unit initially with 60 mg per day together with pyridostigmine up to 540 mg per day. In April 1982 he was discharged with only slight myasthenic symptoms, but the following day he was readmitted with acute myocardial infarction. During hospitalisation he suffered a second acute myocardial infarction from which he also recovered without serious complications. The patient gradually deteriorated despite treatment with pyridostigmine 300 mg per day and prednisone 15 mg per day. He had some degree of respiratory embarrassment, and among other complications was treated for an abscess of the lung. Talking and swallowing became difficult, and he needed a collar to support his neck.

In October 1982 transsternal thytenomy was performed and surprisingly 65 g of apparently thymic tissue was removed. Macroscopic findings: the tissue fragments measured 7 × 7 × 3 cm, 4-5 × 1 × 1 cm and 3 × 2 × 1 cm (three parts) and weighed 65 g. The tissue was soft, yellow and lobulated. A thin capsule covered almost the entire tissue. On cross-section it appeared to be mature adipose tissue divided into lobes by strands. The impression was that of a lipoma. Microscopically, the resected material was mature adipose tissue containing small nodules of thymic tissue with Hassal’s corpuscles. This tissue composition is characteristic of a thymolipoma. During the next 2 months the patient made a remarkable recovery. From November 1982 until June 1983 he was not troubled by signs or symptoms of myasthenia gravis, and, except for a small dose of prednisone (10–15 mg per day) no medication was needed. But in June 1983 he again presented with slight myasthenic symptoms from the face, jaw and sphincters which were easily controlled by methylprednisolone 30 mg four times a day.

Tumours of the thymus are uncommon. Thymolipoma, which is a benign tumour composed of thymic tissue and mature adipose elements, have constituted 2–9% of all thymic tumours.\(^6\) The nature of the tumour is unknown. The two most common clinical symptoms are dyspnoea and cough, but in about 50% of the reported cases thymolipomas were asymptomatic and were detected with routine chest radiography.\(^7\) The radiological appearance of the thymolipoma (or mediastinal lipomatosis of non-thymic origin) is variable.\(^8\) More than 40% of thymolipoma appeared as a cardiomegaly under X-ray examination.\(^6\) In all four cases of thymolipomas in association with myasthenia gravis, the thymolipomas were not detected by usual chest radiographs and conventional tomograms of the mediastinum. Computer tomography of the anterior mediastinum revealed a widening at the level of the great vessels in the case described by Otto et al.\(^9\) and might have revealed the tumours in the other three cases if it had been performed.\(^6\)\(^8\) Removal of the thymolipoma was in our patient followed by complete remission from myasthenic symptoms for six months when a slight relapse occurred. The patient reported by Olanow et al.\(^3\) made a complete recovery, whereas the patient reported by Otto et al.\(^4\) relapsed. The patient reported by Reintgen et al.\(^5\) "showed dramatic improvement" three months following thytenomy. In all four patients the tumour was totally removed.

I thank Dr Stein Poulsen for drawing my attention to the correct diagnosis in this case.

### References


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### Spinal blastomycosis—case report

Sir: Blastomyces is a fungal infection primarily involving the lungs, haematogenous spread may secondarily involve skin and rarely brain.\(^1\) A spinal cord compression syndrome secondary to blastomyosisis of the vertebral bodies has been described\(^2\)\(^3\) but is particularly rare in this country.

We report the case of a previously fit 52-year-old Greek male who began to complain of interscapular pain 16 months prior to admission in June 1981 to the National Hospital for Nervous Diseases, London. Plain spinal radiographs at that time taken by the patient’s brother, who is a radiologist, were reported normal. The pain spontaneously improved and then worsened and five months prior to admission radiography revealed partial collapse of the 4th and 5th dorsal vertebrae with destruction centred on the underlying disc space. The patient had a strongly positive Mantoux test and raised sedimentation rate. The patient was treated with antituberculous therapy without biopsy and presented to us with a three months’ history of progressive cord compression. For the last fifteen years the patient had lived in Africa, particularly in Nigeria, Libya and...