Myeloma of the oesophagus

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SYNOPSIS A case of myeloma of the oesophagus is presented. A search of the literature has revealed no other. The tumour was treated by local resection and postoperative treatment with cyclophosphamide was attempted. The patient died 14 months after operation from a massive myocardial infarct and at necropsy no tumour was found in any organ.

This case is presented because of its extreme rarity. An exhaustive search of the literature has revealed no other case report of a myeloma of the oesophagus.

A 59-year-old woman of Austrian Jewish extraction was admitted to Hammersmith Hospital on 5 November 1968. She gave a history of intermittent difficulty in swallowing solid foods, becoming progressively worse, for four to five months. She had lost 12 lb in weight in the previous four weeks. There was no history suggestive of hiatus hernia.

Her previous history included a myocardial infarct in 1961 and 14 years of diabetes controlled by insulin.

Physical examination was normal.

A full blood count was normal, with an ESR of 72 mm/hour. Liver function tests were normal except for a nucleotidase of 32 IU (normal 2-17). Plasma electrophoresis showed an albumin level of 2.9 g/100 ml (normal 3.3-5) and raised α_1 and α_2 globulins. No paraproteins were demonstrated. A chest radiograph was normal. A barium swallow showed a tumour of the lower oesophagus which had a smooth outline and appeared to indent the oesophagus from one side only. Although the most likely diagnosis was carcinoma of the oesophagus, the possibility of the lesion being a leiomyoma was discussed.

At operation on 12 November, the liver was normal. A tumour, macroscopically like a carcinoma, was confined to the lower thoracic and abdominal oesophagus.

The cardia of the stomach and the oesophagus up to the azygos arch were resected together with a few small lymph nodes.

At the end of the operation two nodules were found in the right lung and were presumed to be

metastases. In view of the apparently poor prognosis these were not biopsied.

Pathology

The specimen consisted of 10 cm of oesophagus and 3 cm of stomach. The lower 7 cm of oesophagus was surrounded by firm white tumour protruding into the lumen. The cut surface of the tumour apparently infiltrated the full thickness of the oesophageal wall which was 1.7 cm at its maximum. The tumour was not weighed, but from its dimensions it was calculated to weigh at least 60 g. The tumour encroached about 0.5 cm into the stomach. Ten lymph nodes were sectioned.

There was a fairly uniform appearance throughout the tumour. It extended to the oesophageal epithelium which, apart from one small area of ulceration, was intact. The oesophageal muscle was infiltrated and in parts destroyed, and tumour cells were present in the surrounding connective tissue. The main bulk of the tumour was confined to the oesophagus but groups of tumour cells were infiltrating into the proximal 0.5 cm of the submucosa of the stomach. The mucosal surface of the stomach was intact.

The tumour was composed of variable lymphoid cells, some poorly differentiated and unclassifiable, some showing differentiation towards plasma cells, and some resembling mature plasma cells. In sections stained with methyl green and pyronin (Unna Pappenheim) over 50% of the cells showed some degree of pyroninophilia.

All 10 lymph nodes were free from tumour.

The postoperative course of the patient was uneventful.

The ESR continued to rise from 85 mm/hour on 21 November to 107 mm/hour on 4 December.

Tests for urinary Bence-Jones proteose were

negative, as were tests for serum paraproteins. The serum albumin continued at a low level and the α and β_1 globulins at a high level throughout her time in hospital. Immunoglobulins were normal except for γ M which was 210 mg/100 ml (normal range 50-150). A skeletal survey showed no bony deposits. Sternal marrow was normal. Liver function tests and prothrombin time were normal.

The patient was discharged home on 5 December and readmitted on 2 January 1969 for two days. She was feeling well and gaining weight. Physical examination revealed no abnormality.

Investigations seven weeks after operation gave the ESR as 38 mm/hour. The LDH, alkaline phosphatase, and nucleotidase were normal. Serum albumin was 2.7 g/100 ml, globulins 4.3 g/100 ml (α and β_1 raised). No Bence-Jones protein was detected in serum or urine. A skeletal survey showed no bony metastases. Chest radiographs showed a few opacities which tomography indicated were likely to be metastases.

It was decided to treat the patient with cyclophosphamide, but, as she took this only occasionally, the total dose taken is not known.

In February 1970 she died from a massive myocardial infarct.

Postmortem examination at Singleton Hospital, Swansea (Dr Williams), revealed no evidence of tumour in any organ.

Discussion

There are several points of interest in this patient. First, no case of myeloma of the oesophagus has been reported. Only 29 cases of solitary gastric

myeloma are reported and only two of these involved the upper end of the stomach (Fraser, Schuh, and Mullen, 1966; Brook, Floyd, and Bliss, 1965; Line and Lewis, 1969).

The absence of Bence-Jones protein and paraproteins is not remarkable, for 20% of patients with extramedullary plasmacytomas have neither (Hobbs, 1969).

It is interesting that the nodules in the right lung, which at operation looked like tumour metastases, had disappeared by the time she died two years later. It is well known that metastases from certain tumours may regress for a considerable time if the primary is removed, but this has never been recorded in cases of plasmacytoma. It is possible that the cyclophosphamide which she took destroyed these nodules, for they might be expected to be highly sensitive to the drug. However, she took it only sporadically and it was not likely to have had a maximal effect. There was no histological evidence that these nodules were in fact myeloma, but if they had been benign lesions it is unlikely that such tumour-like deposits would have disappeared by the time of the necropsy.

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