Section of Surgery

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Short Papers

A Review of 17 Cases of Medullary Carcinoma of the Thyroid Gland

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Cancer of the thyroid gland is a rare disease and this communication discusses a rare variant of it.

In 1959 Hazard *et al.*, in reassessing cases of anaplastic or undifferentiated thyroid tumours, established medullary carcinoma as a clinico-pathological entity. Reviews of thyroid cancer are published currently without reference to it.

Table 1
Histological type of 222 cases of cancer of the thyroid: Hammersmith Hospital and King's College Hospital 1950–66

	No. of cases			
	Female	*Male*	*Total*	*%*
Follicular	40	14	54	24·3
Papillary	55	26	81	36·5
Medullary	7	8	15	6·8
Anaplastic	48	10	58	26·1
Miscellaneous	12	2	14	6·3
Total	162	60	222	

Table 2
Swelling of thyroid gland in 17 cases of medullary carcinoma

No. of cases	*Duration of swelling*
1	Occult
1	On admission
8	Less than 5 years
7	5 years or more ●

● 5, 12, 15, 18, 30 and 31 years and 'years'

In 1966 we reviewed 222 cases of cancer of the thyroid presenting at Hammersmith Hospital and King's College Hospital over sixteen years (Table 1). Since a number of these were referred from other centres for radiotherapy following biopsy or some kind of surgical excision elsewhere, this represents a highly selected group. In this series there were 15 cases of medullary carcinoma giving an incidence of 6·8%. We have added another 2 cases, bringing the total number of cases of medullary carcinoma to 17.

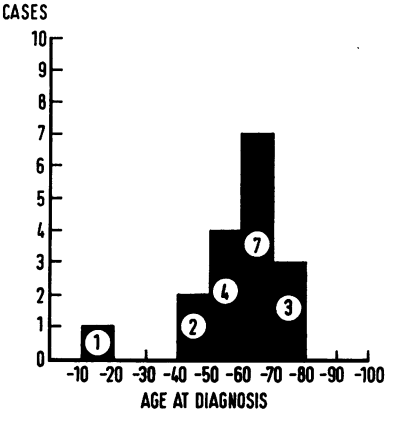

Fig 1 *Age distribution in 17 cases of medullary carcinoma of the thyroid gland*

Clinical details: The sex incidence is equal (Table 1). This is unlike other carcinomas of the thyroid in which the incidence is predominantly female. The age distribution shows a maximum incidence in the seventh decade (Fig 1). All these patients had swelling of the thyroid gland except one in whom the tumour was an incidental post-mortem finding. Many of these swellings were of long standing and 2 were said to show the changes of nodular goitre (Table 2).

Morbid anatomy: Before this tumour was described 14 of these cases were classified as anaplastic or undifferentiated. The histological appearances are characterized by the presence of amyloid in the stroma of the tumour which is composed of sheets and nests of eosinophilic granular cells possibly parafollicular in origin (Williams 1966*a*) (Fig 2). It shows little tendency to encapsulation.

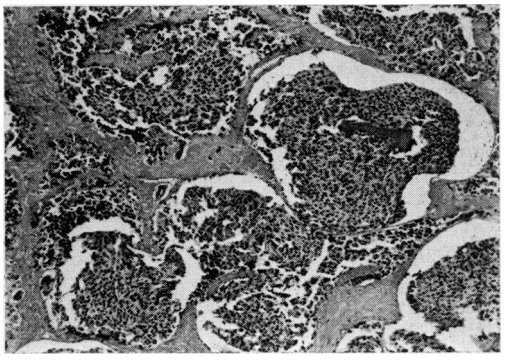

Fig 2 *Medullary carcinoma of the thyroid. Note the strands of amyloid and arrangement of tumour cells in nests.* ×100

Another marked feature is the involvement of cervical lymph nodes and 14 patients developed lymph node deposits. Distal spread may occur in skin and subcutaneous tissues, distal nodes, liver, lungs and bones.

Response to treatment: This tumour is different from other types of thyroid cancer in its lack of response to various methods of treatment:
(1) 11 patients received deep X-ray therapy and one only showed an appreciable response.
(2) 10 patients were given ^{131}I and in none did the tumour or its secondaries show uptake.
(3) 9 patients received thyroxine and none showed evidence of hormone dependence.

The methods of surgical treatment used were variable and it is difficult to draw conclusions from survival times following the various operations performed in only 13 patients. Accepting the common aim of removing tumour and involved nodes it might appear that the presence of cervical node deposits at the time of operation would affect prognosis. Nine patients had local cervical nodes involved at the time of operation and they had a mean survival time of five years. Three patients had no node deposits at the time of operation: their mean survival time was nine years. This difference is not significant ($0{\cdot}2 < P < 0{\cdot}5$).

Duration of survival: Of 16 patients, 4 survivors are alive at four, seven, seven and seventeen years after the onset of symptoms. There have been 12 deaths, 10 from thyroid cancer; their mean survival time was five years, the range being from five months to twenty-three years.

This disease has a potential for killing quickly or slowly and its rate of growth is intermediate between anaplastic carcinoma and differentiated carcinoma. Total thyroidectomy with removal of involved nodes and tissues would seem a reasonable form of treatment.

Table 3
Duration of diarrhœa in 5 patients with widespread medullary carcinoma of the thyroid

Patient	*Sex*	*Duration of diarrhœa*	*No. of motions per day*
C B	F	12 months	
E S ●	M	2 years	6
P L	M	12 years	3
M K	M	15 years	4
J L	M	'Life'	8

● Improved after removal of primary tumour

Williams (1965) pointed out an association between this type of cancer and the occurrence of neuromas and phæochromocytomas. This small and retrospective survey includes one patient with multiple mucocutaneous neuromas and one other possible case, not histologically proven. Two of our patients were found to have raised vanillyl mandelic acid levels in blood and urine. This suggested the possibility of excess noradrenaline and adrenaline secretion by phæochromocytomas. At autopsy in one patient (M K) we found bilateral phæochromocytomas in his adrenal glands. Unfortunately the second patient was lost to autopsy.

Williams (1966*b*) has also pointed out that a significant number of these patients have diarrhœa. It is suggested that this is a manifestation of serotonin-kinin secretion by the tumour tissue. Of these 17 patients, 5 suffered persistently loose motions of abnormal frequency. Table 3 shows the duration of diarrhœa from the time of onset to hospitalization and the number of motions per day. No adequate cause was established in any case. All had widespread tumour and one improved after operation. Two patients remain alive and we hope to estimate their serotonin levels; 2 died before this association was suggested. The third who died had normal serotonin levels in his urine and had been aware of a thyroid swelling for thirty-one years; he had an 89-year-old mother who had a goitre for as long as he could remember; she suffered also from longstanding diarrhœa.

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