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Case Report

Papillary Carcinoma of the Thyroid Gland and Atresia of the Common Bile Ducts of a Five Year Old Boy

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Summary. We present a case of papillary carcinoma of the thyroid gland with pulmonary metastases in a 5 year old boy. The child also suffered from atresia of the gallbladder and the common bile ducts with biliary cirrhosis of the liver and died from hepatic insufficiency. Possible correlations between childhood thyroid carcinoma and congenital malformations are discussed.

Key words: Thyroid carcinoma – Thyroid gland hypoplasia – Bile ducts atresia.

Introduction

Although the incidence of thyroid gland carcinoma has increased in the past few years, it is still a relatively rare disease in childhood. Winship and Roswell found 562 cases of children up to 15 years old with thyroid gland carcinoma in the literature up to 1962. Eighty percent of these cases were reported from the USA.

In this report we shall discuss the case of a metastasing thyroid gland carcinoma in a 5 years old male child with congenital gallbladder atresia.

Case Report

The boy was the fourth child born to a 40 years old woman after a normal pregnancy. Her three other children were healthy. After a normal birth the child became jaundiced which led to its hospitalization after 7 weeks. Laboratory results indicated gallbladder atresia and mild liver cirrhosis which were supported by the findings of a liver biopsy and scintigraphy. A total lack of extrahepatic bile ducts and mild cirrhosis was observed at laparotomy. Due to these findings a hepato-jejunostomy was performed on the boy when 16 months old.

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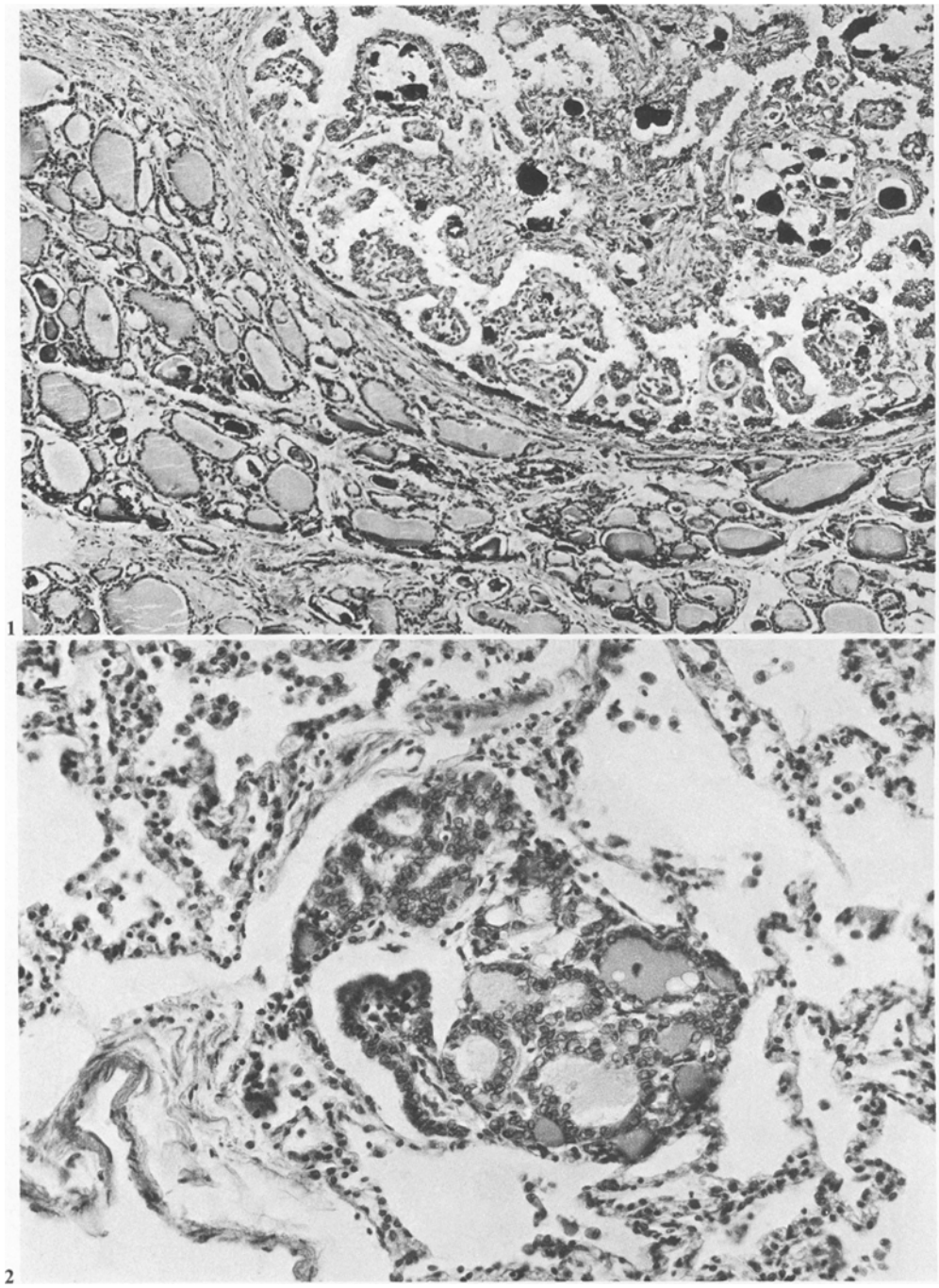

Fig. 1. Papillary carcinoma of the thyroid gland of a five year old boy. Note difference in appearance between carcinoma and thyroid gland and the presence of an ill-defined capsule

Fig. 2. Metastasis of the thyroid gland carcinoma in the lung

During the next three years the child developed normally at first but was then found to have increasing liver insufficiency which eventually led to his death at 5 years of age. No hormonal checks were carried out during the boy's life.

Post mortem examination revealed a fine nodular liver cirrhosis with considerable pseudobile duct proliferation, extensive bile stasis and freshly developed liver cell necrosis. In addition a cholaemic nephrosis and 4 litres of ascitic fluid was found. The operative connection of the jejunum to a proximal portion of the main hepatic duct was intact.

The thyroid gland was only 1 cm in diameter. In the left half a pea-sized, grey-white tumor was found. Histologically the tumor proved to be a papillary and follicular carcinoma. Except for slight, striped fibrosis of the interstitium, the rest of the thyroid gland showed no anomalities histologically (Fig. 1). Metastases were found on microscopical examination of the lungs (Fig. 2). All other organs of the child, including the other endocrine glands, especially the pituitary gland and the adrenal gland were found to be normal.

It was not possible to conduct a chromosomal study, the boy having being dead for more than 12 hours at the time of the post-mortem examination.

Discussion

Meissner (1968) showed that the occurence of thyroid gland carcinoma in childhood occured twice as often in males as in females. The examinations which Winship and Roswell did up to 1962 showed that the majority of the young patients lay within the 4 months–15 years age range, with an average age of 9.6 years. In their report 12 of the children had been born with a thyroid tumor which was later shown to be malignant. Lymph node metastases and sometimes metastases to the lungs and skeleton are often the first diagnostic signs of the illness (Schmid, 1972).

In this case the thyroid carcinoma was discovered during autopsy with no previous suspision of its presence. Later, in a histological examination, metastases were found by change in the lung sections.

A correlation between X-ray therapy to the throat of babies and infants and the subsequent occurence of thyroid gland carcinoma is very strong (Dufty and Fitzgerald, 1950; Baron, 1958; Hempelmann, 1967). Winship and Roswell (1962) quote a figure that 80% of the cases they studied with thyroid gland carcinoma had previously received X-ray therapy. X-ray therapy was widely used in the USA at one time for the treatment of such illnesses as thymic hyperplasia, enlarged tonsils etc. Children with thyroid gland carcinoma studied in Europe have a much lower percentage history of X-ray therapy (Schmid, 1972). The interval between X-ray therapy and the appearence of carcinoma was approx. 8.7 years (Winship and Roswell, 1962).

In the present case there was no history of X-ray therapy. The cause of this tumor may be linked to a congenital abnormality of the thyroid gland (thyroid gland hypoplasia) as gallbladder atresia was also present. There is little information in the literature to suggest a connection between congenital malformations and malignant tumors in childhood. However, some tumors are connected with malformations, such as leukaemia and Down's syndrom, leukaemia and osteogenesis imperfecta, nephroblastoma and renal abnormalities. As far as the simultaneous occurence of thyroid gland carcinoma and congenitalimalformations is concerned, only one case of thyroid gland carcinoma and congenital malformation of the heart has been reported by Winship and Roswell (1962). The combination of thyroid gland carcinoma and gallbladder atresia is surprising

as the latter disorder is also relatively rare. The cause of gallbladder and biliary duct atresia is still under discussion with the possibility of an infection or developmental disturbance being favoured (Flegel, 1950).

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