CASE OF CUSHING’S SYNDROME DUE TO AN ADRENAL TUMOUR DESCRIBED IN 1914 BY DR. LUCIEN DEDICHEN OF KRISTIANIA (OSLO)

by

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In a paper read before the Norwegian Medical Society in Kristiania (Oslo) on 4 November 1914, Dr. L. Dedichen described a case which possessed all the characteristic features of what was later known as Cushing’s Syndrome. The author explained all the symptoms as being due to hyperfunction of an adrenal tumour, which was found on autopsy. The paper was published in Norwegian, without any summary in English, and therefore escaped the notice of writers on medical history and also of Cushing himself, when he reviewed the literature on this subject in 1932. Dedichen’s case is noteworthy also because there is a complete autopsy report by the noted pathologist Francis Harbitz, with microscopical description of all the principal organs, including the hypophysis. The laboratory studies included glucose tolerance tests, which were not very commonly performed in the medical clinics at that time.

I have a special interest in the case, as I saw the patient when I served as an intern on the staff of the University Medical Clinic B, where the patient was hospitalized.

According to Soffer several cases of precocious physical development associated with adrenal tumours especially in young girls (3–4 years old) had been reported since 1803, and, as Soffer states, there was gradual recognition of the combinations of the adrenogenital syndrome with those additional metabolic disturbances later characterized as Cushing’s syndrome.

Cushing’s first patient, observed by him in 1910, had passed in and out of hospitals without any definite diagnosis having been made. This patient was still alive in 1932 and there was then no pathological proof of disease in the hypophysis. The final report of Dr. Turney’s case, described clinically by him in 1913, was given by Parkes Weber in 1926, twelve years after the finding of an enlarged left suprarenal gland on autopsy in 1914, but not reported at that time. The case of Dr. Anderson was published in 1914, almost simultaneously with that of Dr. Dedichen, in a journal probably not available in Norway at that time. Anderson found a pea-sized adenoma of the adrenal gland and a millet-sized tumour in the anterior hypophysis.

Dr. Dedichen’s report, which was part of a paper dealing with several endocrine disorders, is very short and gives only the essential data.
The case was that of a 23 year old married woman with one child (aged 4). She had developed amenorrhea and increasing obesity accompanied by headache, loss of hair of the head, but otherwise with hirsutism, especially of the face. There was found abnormal pigmentation, hypertension (215 mm. systolic pressure), proteinuria, polyglobulia with tendency to subcutaneous hemorrhages, lowered glucose tolerance and occasional glucosuria. The tolerance test included blood sugar determinations by the recently described micromethod of Bang. Dedichen does not, however, give any details in his short account. He does not mention the purple striae on the abdomen and the buttocks, but they are described in the status praesens, taken up on the patient's arrival at the clinic, and can be seen in the picture (Fig. 1b). The eyegrounds showed 'retinitis albuminurica' and blurring of the disk margins. The liver and the spleen were much enlarged and a tumour was felt below the liver, which was interpreted as an enlarged right kidney. There was also considerable hypertrophy of the heart.

Dedichen discussed these findings and concluded that an adenoma of the adrenal cortex was the most probable diagnosis. He mentioned that the experience of recent years made it probable that, in adults, hypernephroma might be associated with the symptoms that his patient had developed, namely, amenorrhoea, adiposity and abnormal hair growth.

The autopsy performed a few hours after death (Professor Harbitz) revealed a typical rightsided hypernephroma, while the hypophysis was normal. Dedichen concluded his short report by mentioning that those hypernephromas which might be associated with the above-mentioned symptoms were real adenomas; later they might become malignant and they originated from the adrenal cortex itself and not from ruptured parts of the interrenal system. The so-called Grawitz tumours on the other hand were associated with symptoms of hyperfunction of the chromaffine system.

Here ends the short report of Dedichen without any further presentation of the pathological findings. These are recorded in the autopsy report of Harbitz (autopsy No. 113/1914) which contains a detailed and exact description of the organs and includes a careful microscopical examination. The adrenal tumour was manhead-sized with a smooth surface and had the typical appearance of a hypernephroma. The much enlarged liver was full of metastatic, yellow brown tumours. Similar tumours were also found in the lungs. The hypophysis was normal macroscopically and weighed 0.4 g.; the microscopical examination showed normal structure.

In the microscopical description of the hypernephroma one recognizes the characteristic suprarenal structure: large, clear, protoplasma rich cells, arranged in narrow parallel rows, sometimes collected in small heaps, but the nuclei were badly stained and there were large areas of degeneration. This picture was also seen in the metastases in the liver and the spleen.

It is hoped, that this short account of a pioneer clinical observation will prevent Dr. Lucien Dedichen's name from being entirely forgotten and also be a witness of the high standard of Norwegian clinical medicine in the early part of this century.
Dr. Dedichen’s case of Cushing’s Syndrome. Showing the patient before and two years after the development of symptoms.
A Case of Cushing's Syndrome

REFERENCES