ROKITANSKY—KUSTER—HAUSER SYNDROME

(A Case Report)

by

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Introduction

Rokitansky-Kuster-Hauser syndrome with its multiple skeletal, genital and urinary tract abnormalies is presented. The plausible aetiological factor is discussed.

Case Report

S., aged 18, presented with pain and lump in abdomen of 3 months duration, associated with primary amenorrhoea. The only relevant past history was that she was born preterm at 7½ months gestation to an elderly primigravida of 37 years age following 23 years of infertility and she was the only child. Imperforate anus detected at birth was surgically corrected the same day. She was only 4 feet 6″ in height with webbed neck—She had no cubitus valgus, but hypothenar eminence was absent. The secondary sex characters were not well developed. A mobile, non-tender well defined mass 10 x 8 cm was felt in lower abdomen. The vagina appeared to be absent.

X-ray chest showed normal lungs, and kyphoscoliosis of thoracic spine. X-ray neck revealed fusion of 5th, 6th and 7th cervical vertebrae. X-ray pelvis showed absent ala on right side.

Laparoscopic study revealed a mass surrounded by omental adhesions, and because of adhesions, neither the nature of this mass, nor the genital organs could be made out. Absence of vagina was confirmed under anaesthesia.

The chromosomal study revealed the presence of barr bodies. Non functioning of right kidney was detected on IVP—Left kidney appeared normal.

Retrograde cystoureterogram showed marked right hydroureter (Photograph I).

At laparotomy, the mobile mass turned out to be a right sided ovarian cyst surrounded by omental adhesions. The fallopian tube was normal, but the uterus which was unicornuate and rudimentary ended in a fibrotic band without a cervix. On the left side, neither the ovary nor the mullerian tissue could be seen. Right sided hysterectomy with ovariotomy was carried out. This was followed by right nephroureterectomy. Her post-operative period remained uneventful.

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See Fig. on Art Paper VII