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CASE REPORTS

OMPHALOCELE

J. E. LEDDY, F.R.C.S.[C.], F.R.C.S.(Edin.)
and A. E. BUCKWOLD, M.D., D.C.H.,
Saskatoon, Sask.

AT THE FOURTH WEEK of intrauterine life the greater part of the midgut is extruded from the main abdominal cavity by the rapid increase in size of the liver and other organs and the midgut thus lies in the umbilical cord. When the midgut fails to return to the abdomen the condition known as omphalocele results and normal rotation of the gut cannot occur. Specht and Shryock[1] state that "the most commonly accepted explanation is that there exists a disproportion between the abdominal viscera and the abdominal cavity. This disproportion usually results from a retarded development of the abdominal parietes".

It is generally accepted that surgical repair should be carried out on the first day of life preferably a few hours after birth. Conservative treatment by application of dusting powder and alcohol compresses followed by strapping has apparently occasionally been successful according to Gross and Blodgett.[2] But almost all cases die within a few days unless operation is performed. Death results from rupture of the sac and evisceration or from infection and peritonitis.

This case is reported for the following reasons: (1) Because it represents survival of a case of omphalocele with most of the liver in the sac, first operated on forty-three days after birth. (2) The development of bilateral inguinal herniæ immediately after the first operation providing extra room for the viscera contributed greatly to the survival of the child. (3) Repair of the ventral hernia when the child was 10½ months of age was followed by a marked increase in the size of the inguinal herniæ. This release mechanism probably ensured the success of the ventral hernia repair.

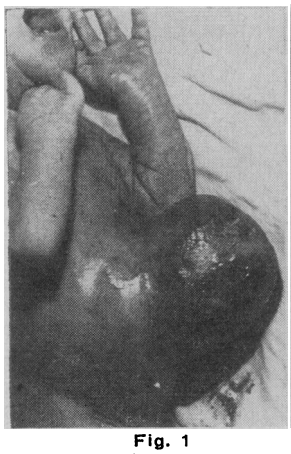

Fig. 1

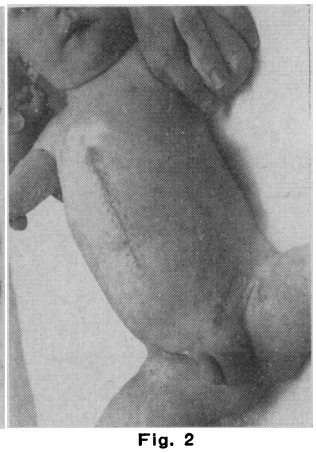

Fig. 2

Fig. 1.—Forty-three days after birth. Fig. 2.—Age 1 year.

L.R.W., a five weeks' old male infant was admitted to St. Paul's Hospital, Saskatoon, in April 1949 for repair of an omphalocele. The infant was full term normal delivery. Birth weight was six pounds nine ounces. The pregnancy had been uneventful and the family history negative. A large omphalocele covered by a shiny translucent membrane through which loops of bowel were visible was present at birth. The omphalocele was treated conservatively and although the child did not gain weight his general condition was fair.

On admission to hospital the infant was poorly nourished, weighing six pounds one ounce. The main finding on physical examination was a large omphalocele 9 cm. in diameter with a maximum protrusion above the anterior abdominal wall of 7 cm. The umbilical scar was evident at the centre of the protrusion. Healthy skin extended beyond the edges of the hernial sac. The hernial sac was covered by a thin reddish membrane on top of which there was a fine film of purulent exudate. The recti were markedly separated. Other abnormalities noted were: an abnormally high hard palate; the right fifth finger was deformed at the metacarpo-phalangeal joint; the right testicle was ectopic and a hydrocele was present on the left side.

First operation.—On the fifth day after admission (43rd day after birth) under open ether the first stage of the repair of the omphalocele was performed by the late Dr. R. H. Macdonald. At operation the transverse colon, part of the stomach, small bowel and practically the whole of the liver were noted in the sac. These organs were returned to the abdominal cavity. However it was impossible to approximate the recti. Without undercutting the edges, the skin was brought together with mattress wire sutures.

The microscopic description of the hernial sac is as follows: "Much of the cutaneous surface is ulcerated and covered by a thick slough of purulent exudate and necrotic tissue debris. Beneath this there is an exuberant granulation tissue proliferation with much chronic inflammatory reaction. The corium is scarred and similarly inflamed with many new vascular channels. The inflammatory reaction even spreads into the adherent adjacent peritoneum in which there is much fresh hæmorrhage".

Immediately following operation both limbs and the lower abdomen became markedly cyanosed. Heparin was started and the limbs elevated. Cyanosis disappeared in about forty-five minutes but a pitting œdema of the thighs and legs persisted for six days. A stormy postoperative course followed. With oxygen, intravenous fluids, antibiotic therapy and a gradual resumption of feedings the general condition of the infant improved slowly. The improvement was aided by the development of bilateral inguinal herniæ on the fifth postoperative day. By August 1949 his general condition was good. He had two large reducible inguinal herniæ which were controlled by trusses. The patient was discharged to return later for repair of the ventral hernia.

Second operation.—January 27, 1950, age 10½ months, repair of the ventral hernia by one of us (J.E.L.). Under ether anæsthesia, a vertical elliptical incision was made, encircling the hernia in the epigastric region. The viscera were covered by peritoneum and skin only. The anterior and posterior rectus sheaths and peritoneum were identified on both sides and repair was commenced. The peritoneum and posterior sheaths were brought together

in the midline with No. 1 chromic catgut interrupted sutures. The anterior sheath was sutured with interrupted 00 black silk sutures. Vertical Mattress 000 silk sutures were inserted for skin closure.

The postoperative course was generally good but it was noted that immediately after the operation the bilateral herniæ increased greatly in size and that they contained bowel.

Third operation.—February 18. A large indirect right inguinal hernia with a thick-walled sac was isolated. This was repaired in standard fashion. It was noted that the testis was lying in an ectopic position in the subcutaneous tissue just below the inguinal ring.

Fourth operation.—February 24. A large indirect left inguinal hernia was repaired. A large hydrocele was also repaired. On discharge from hospital two weeks later all wounds were healing nicely. Six months later the child was seen again and the repair of the omphalocele was considered to be quite satisfactory.

SUMMARY

A case of omphalocele, containing almost all the liver, is reported. This case was operated on 43 days after birth. Skin coverage of the viscera was obtained at the first operation and 10½ months later repair of the ventral hernia was completed. The rôle of the inguinal herniæ in providing much needed room for the viscera is stressed.

REFERENCES

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605 Canada Bldg.

INTRAPLEURAL THYMIC TUMOUR IN MYASTHENIA GRAVIS*

J. A. PELLER, M.D. and
H. J. BARRIE, B.M., B.Ch., *Toronto*

THE PERSISTENT or enlarged thymus associated with myasthenia gravis is usually situated in the superior mediastinum and projects up into the neck, or more commonly down into the anterior mediastinum, depending on the degree of enlargement. Because of the recent interest in thymectomy as a treatment for myasthenia gravis and because no similar cases have been reported, we are presenting a case in which the enlarged thymus was suspended in the right pleural cavity.

SUMMARY OF CLINICAL HISTORY

This 46 year old farmer was in excellent health until three months before death when he developed weakness and clumsiness of the hands. A few weeks later, the weakness spread to his arms and shoulders. One month before death, the strength in his hands returned to normal, his arms improved, but his shoulders remained unchanged. About this time he noted difficulty in holding his head erect, and progressive difficulty in swallowing. Two weeks later his face felt "stiff and hidebound"; mucus collected in his throat because of impaired coughing. In the week before admission to hospital, his voice became nasal and he had occasional transitory diplopia when looking to the left or right. There was no family history of musculo-skeletal disease.

Examination in hospital three days before death, showed a well built, sturdy man. The eye movements were full and there was no diplopia. There was a bilateral symmetrical upper and lower facial paresis. The pharyngeal reflexes were present but there was slight weakness of elevation of the left side of the soft palate. The voice had a definite nasal character. The sternomastoids were equally weak, but the trapezii were strong. The tongue could be moved from side to side, but could not be protruded beyond the teeth. He was able to shrug his shoulders, but there was marked symmetrical weakness of all other movements of the shoulders and elbows and upper part of the legs though distal power was preserved in all limbs. There was only faint excursion of the thorax, and fluoroscopy showed normal diaphragmatic movement. The rectus abdominis was very weak. There was no wasting, pseudohypertrophy, fibrillation, or disturbance of tone. The blood and cerebrospinal fluid were normal. A routine miniature chest x-ray taken on admission, but not read until after his death, showed an opacity in the right cardiophrenic angle.

A test dose of 0.5 mgm. of prostigmine was injected hypodermically. In about 15 minutes there was a slight lessening of the nasal quality of his voice, and a slight improvement in his ability to cough. The lower facial muscles were slightly stronger. There was no improvement of the upper limbs, sterno-mastoids, or muscles of the tongue. He was subsequently given four hypodermic injections of prostigmine, each being followed by the same slight transient improvement. About six hours after the last injection, he was seen by a nurse and was resting quietly. Three minutes later he was found dead.

SUMMARY OF AUTOPSY FINDINGS

There was no cyanosis of the body. An oval mass weighing 55 gm. and measuring 9 x 6 x 3 cm. lay in the right pleural cavity between the medial aspect of the right lung and the right lateral surface of the heart, approximately at the level of the minor fissure. The mass was suspended from the superior mediastinum by a tubular reflection of parietal pleura resembling a mesentery and containing an artery, two veins, and some fatty areolar tissue (Fig. 1). The tumour was pinkish-grey, soft, cystic and enclosed by a thin fibrous capsule. In the fixed specimen, the cut surface showed a granular, follicular parenchyma traversed by thin fibrous septa. The lungs were heavy, weighing 750 gm. each. Petæchial hæmorrhages and areas of bronchopneumonic consolidation were scattered through the lower lobes. The spleen was enlarged, weighing 325 gm. The cut surface revealed normal splenic tissue. The cerebral hemispheres were œdematous. A small grey nodule, 5 mm. in diameter, projected into the fourth ventricle from the superior medullary velum.

Microscopically, the parenchyma of the intrapleural tumour consisted of sheets and follicles of lymphocytes divided into lobules by fine fibrous trabeculæ. Large pale epithelial reticular cells and well-developed Hassall's corpuscles were distributed evenly through the lymphoid tissue. The numerous capillaries and venules were distended by red blood cells. The appearance was that of a uniform benign hyperplasia of the thymus. Sections of the affected muscles showed small perivascular accumulations of lymphocytes. The small nodule projecting into the fourth ventricle was an astrocytoma. The cause of death was not determined.

DISCUSSION

The clinical diagnosis of the case was difficult. The facio-scapulo-humeral distribution of weakness, the absence of myasthenic reactions and

*From the Department of Pathology, University of Toronto