Appendicular Tumors

W. E. HIRTLE, B.Sc., '41

THE rareness of this entity must be stressed at the outset. Though tumors of the appendix are few in number compared to all appendicular pathology they should, however, be considered in all cases where a diagnosis of chronic appendicitis is made pre-operatively.

Several types of tumors must be considered including: carcinoma, neuroma, myxoma, angioma and mucocoele.

SYMPTOMATOLOGY:

The symptoms are much like those of chronic appendicitis, so much so that this is the usual diagnosis made where tumors of the appendix are present. Pain is the most outstanding symptom. It is usually localized to the right lower quadrant of the abdomen, though some cases have been reported where the pain is referred to the right upper quadrant and even to the epigastrium. The pain is usually present over a period of a year or more and is more recurrent in nature, rather than persistent. Some cases have been reported in which the pain became severe enough to require the administration of morphine. Between attacks there is usually some residual tenderness in the right lower quadrant.

Fever, if present at all, is of low grade, never rising above 100°F. Nausea and vomiting are early symptoms. This is explained by the fact that the appendix becomes distended early, thus bringing about reflex nausea and vomiting. In the case of a cystic appendix this is more marked due to more complete stretching of the appendicular coats.

The blood picture is fairly normal. Typical findings are as follows:

Haemoglobin	70–80%
Red Blood Cells	4,000,000–5,000,000 per cubic millimeter
White Blood Cells	9,000– 10,000 per cubic millimeter

PATHOLOGY

Carcinoma:

Under this heading two types must be considered. There is first of all the "true carcinoma", which is the less important of the two. This type is usually the columnar celled or the adeno-carcinoma. In most cases the lumen of the appendix is partially or wholly blocked and in some cases the serous coat is infiltrated.

The second type of carcinoma is the "carcinoid" tumor of the appendix. It is the more common of the two groups and is not a true carcinoma. However, it resembles a carcinoma microscopically but is essentially benign in its course. It is usually superimposed on a previously inflamed appendix, which becomes thickened and fibrosed. There is generally a hard nodule at the distal third of the appendix which on cross-section presents a yellowish appearance. Microscopic examination of the carcinoid tumor reveals spheroidal and polyhedral cells. These cells are rich in lipoid material and are apparently from the Kultschitsky cells

which lie between the columnar cells of the crypts of Lieberkuhn. Because these cells belong to the chromaffin system the tumor is sometimes called a chromaffinoma. Likewise because the tumor stains by means of silver impregnation, it is also called an argentaffinoma.

Neuroma:

Because of a possible relationship between neuromas and carcinoid tumors, it is better to mention Neuromas at this time. They are thought to arise from the non-medullated nerve fibres of the mucosa and according to Masson, argentaffin cells migrate into the nerve fibres and a neuroma results. Should these cells proliferate, a carcinoid tumor of the appendix is produced.

Polyp:

These are of necessity small in nature and are apparently found in any portion of the appendix. Because they block the lumen they naturally obstruct normal drainage and thus tend towards mucocele. The polyps seem to consist of mucous membrane, with a small portion of the submucosa jutting into the lumen of the appendix.

Myxoma:

Here the appendix is slightly enlarged and has a myxomatous appearance. On microscopic examination the cells show a stellate appearance with some cytoplasmic processes. Unlike true myxomata, the cells contain for the most part single nuclei.

Haemangioma:

This growth is very rare but cases have been reported in which the appendix was noted to have a brownish mottled appearance and on cross section appeared like a dark red sponge. The histology shows venous spaces confined mostly to the muscular and serosa layers. These spaces are lined with endothelial cells and in some cases are filled with these and red blood cells.

Mucocele:

Because of the relatively more frequent occurrence of this type of tumor it will be treated more fully.

The cause is thought to be some obstruction. The causes can be subdivided into (a) extrinsic and (b) intrinsic.

(a) The extrinsic cause resolves itself into adhesions, causing a kinking of the appendix with consequent obliteration of the lumen.

(b) The intrinsic factors are three in number:

1. Concretion.
2. Involution or obliterative process causing occlusion between the secretive area and the base.
3. Specific infections such as: Tuberculosis, Syphilis, Typhoid, and ulcerative inflammatory processes.

Mucocele varies in size from 5-12 c.m. in length and 15m.m. to 8 c.m. in width. The shape is usually oval or tubular though many variations occur.

The origin of the contents of a mucocele is still in the theory stage, some claiming its source from the mucosa and connective tissue while still

others think it due to degeneration in an already atrophic or senile appendiceal wall. The material is usually gelatinous in nature and varies in color from grey to yellow.

The mucosa is usually ulcerated, thinned, or may even be absent. The muscle layer is generally hypertrophied or fibrotic; the serosa may show hyperplasia of blood vessels but later this becomes attenuated.

When there is extension of the gelatinous material through the coats of the appendix, or when the appendix ruptures, it usually sets up a condition in the peritoneal cavity, known as "pseudomyxomata peritonei".

DIAGNOSIS:

The Diagnosis of tumors of the appendix is usually made post-operatively. Conditions which resemble tumors in symptoms and signs are:

1. Appendicitis and appendicular abscess.
2. Tumors of the ilio-caecal region.
3. Tuberculosis and actinomycosis.
4. Retroperitoneal masses, as renal ptosis and psoas abscess.

In women we may add several more conditions:

1. Ovarian cysts and tumors.
2. Hydrosalpinx and unruptured tubal pregnancy.
3. Pedunculated tumors of the uterus.

TREATMENT:

In all cases the treatment is Surgery, even in the cases of psuedomyxomata peritonei.

PROGNOSIS:

The prognosis is unusually good. There are no cases on record where there has been evidence of metastases even in microscopically malignant tumors. Even in cases of pseudomyxomata peritonei the prognosis is fair when the source of the peritoneal involvement has been removed.

BIBLIOGRAPHY

Boyd—Textbook of Pathology, 2nd Edition, p. 543-544.
Doyle, J. C.—Mucocoele of the Appendix.
The Western Journal of Surgery, Obstetrics and Gynecology (Sept. 1939).
Norment, W. B.—Tumors of the Appendix.
Surgery, Gynecology and Obstetrics, Nov. 1932 (Page 590-595).

"No other profession is expected to give its services so freely without fee or reward. This is partly due to the high standard of generosity that long tradition has led the public to expect, and partly to the fact that the doctor renders services that are a necessity to the community, and it is unthinkable that lack of means to pay should deprive any individual of his most pressing need in time of sickness."—*E. Kaye LeFleming.*