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Review Article

Tumors of the Appendix: A Surgical Enigma

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ABSTRACT

Tumors of the appendix are rare. They are usually diagnosed at the time of emergency appendectomy for acute appendicitis. An elaborate histopathological evaluation followed by a detailed staging of the disease dictates the definitive surgical management of these tumors. The paper reviews the pathology and surgical options for appendicular tumors.

Key Words: Tumors, Appendix, Carcinoma, Carcinoid, Diagnosis Treatment

INTRODUCTION

Appendectomy is one of the commonest operations performed in general surgery. The decision to perform surgery is based solely upon clinical criteria supported by laboratory and radiological investigations. However majority of surgeons forget to open the specimen prior to dispatch for histopathological examination. As a result many a time's tumors of appendix are missed intra operatively. Tumors of appendix are peculiar with respect to their biological behavior. The surgeon needs to understand the pathology and natural history of appendicular tumors in order to make a valued judgment regarding the definitive surgical treatment for these malignant tumors. The surgical pathology and treatment of appendicular tumors is presented in this paper.

Pathology:

Tumors of appendix are rare. Traditionally carcinoid tumors are found to be more common than adenocarcinoma of the appendix. [1] However with time, more meticulous evaluation of appendectomy specimens the incidence of carcinoid tumors have been superseded by other tumors usually mucinous adenocarcinoma. [2] Mucinous cystadenocarcinoma are highly malignant tumors of appendix. These are lined by atypical mucinous epithelium with some areas of papillary pattern. Due to obstruction caused by the tumor there is accumulation of mucus giving rise to a mucocele of appendix. [3] This if untreated and allowed to rupture or if accidently ruptured during surgery can lead to the development of pseudomyxoma peritonei syndrome, a condition which has got extremely high morbidity and mortality. Therefore if a surgeon encounters a

mucocele at the time of emergency surgery, great caution and care needs to be exercised while removing such an appendix.

The other types of malignant tumors are primary adenocarcinoma of appendix which are quite rare followed by signet ring type or linitus plastica type of carcinoma. [4-6] Signet ring carcinomas usually have more extensive mucosal involvement, more nuclear atypia and more complex and irregular invasive pattern as compared to mucinous tumors. In majority of cases, these are incidental finding associated with appendectomy. [5,6] Carcinoids are quite commonly encountered in the appendix. [7] Most are associated with an attack of acute appendicitis. Majority are located at the tip of appendix with size usually less than 1 cm. The tumors are firm, yellow and well circumscribed. The tumors located at tip are closely described as 'Bell clapper configuration". Carcinoid tumors exhibit great histological variation and three categories have been described. [7,8] The classic type comprises of solid nests of small monotonous cells with occasional acinar or rosette formation. Mitosis is exceedingly rare and the cells are argentaffin, argyrophillic, and positive for diazo reaction. They are filled with granules containing neuron specific enolase, chromogranin, serotonin and other peptide hormones such as somatostatin, substance p, Glucagon and Peptide Y.

The second type of carcinoid tumor is referred to as a tubular type of adenocarcinoid. It is characterized by gland formation without solid nests. Theses lack mitosis and atypia. On occasion these cells are large, acidophilic, simulating panneth cells. These features are also exhibited by normal "Kulchitsky' cells". The third variant is called mucinous carcinoid or goblet cell carcinoid. Grossly it appears as an area of whitish or mucoid induration of any part of appendix without dilation of the lumen. Like the two previous carcinoid types, it is predominantly a submucous growth. This category of carcinoids exhibits lysosomes positive cells which stain positively for mucicarmine and CEA stains. The granules are also stained positively with argyrophillic stains.

The peculiarity of carcinoid tumors of appendix is that they rarely exhibit the classical carcinoid syndrome. [7,8] As a result majority goes undiagnosed, in subjects who otherwise do not develop acute appendicitis. Patients presenting as carcinoid syndrome usually have liver metastasis.

Surgical implications:

Though elaborate knowledge of the pathology of appendicular tumors is available, yet the surgical application of this knowledge creates a state of confusion in the mind of attending surgeon. Various studies have suggested various surgical options for appendicular neoplasms. However the majority of these series are based upon incidental diagnoses made on post operative specimens. The location of the tumor in the appendix is very important factor while making a decision for definitive treatment. [9] Malignant tumors may at times involve the base of appendix and adjacent cecum. In such cases mesenteric lymphadenopathy may be common accompaniment. It is therefore advisable to open every appendectomy specimen at the time of surgery before commencing closure of the incision. If any suspicious lesion is detected intraoperatively in the specimen, then that warrants further assessment of the cecum, terminal ileum, mesoappendix along with adjacent mesocolon. This would yield additional information to the staging process. Any heroic radical surgery at the time of appendectomy is to be avoided until proper histological and histochemical diagnosis is arrived at. [10] Once the definitive diagnosis is made by histology, a staging of the disease by contrast enhanced

CT scan of the abdomen is essential. This will give additional information of local and distant spread especially to liver. If the liver is spared of metastasis then right radical hemicolectomy would be safest and best option especially in cases of adenocarcinoma. Controversy still surrounds the treatment of appendicular carcinoids and an extensive and radical procedure in non metastatic carcinoid is unwarranted especially when carcinoid tumor is located at tip with normal intervening layers of appendix. A carcinoid less than 2 cm located at the tip with low proliferative index and devoid of angiolymphatic or mesoappendiceal or extra appendicular spread can best be treated by just appendicectomy. [1,11] However if carcinoid happens to be at base then there is always possibility of cecal wall involvement. This accompanied by high mitotic index, mesoappendiceal invasion or a goblet cell adenocarcinoid warrants a radical right hemicolectomy. [1,11] Majority of incidentally found malignant tumors of appendix are picked up at primordial stage where right radical hemicolectomy may suffice. However in the event of metastasis being detected adjuvant chemo may be necessary in select number of cases.

CONCLUSION

Every appendectomy specimen on removal should be cut open and inspected thoroughly prior to closure of incision.

Any suspicious appendicular lesion warrants elaborate histopathological, histochemical and radiological evaluation.

Having diagnosed and staged the malignant lesion definitive radical right hemicolectomy should be performed on an elective basis at the earliest.

No attempt should be made to perform a radical procedure based merely upon suspicion of a gross malignant lesion at the time of emergency appendectomy.

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