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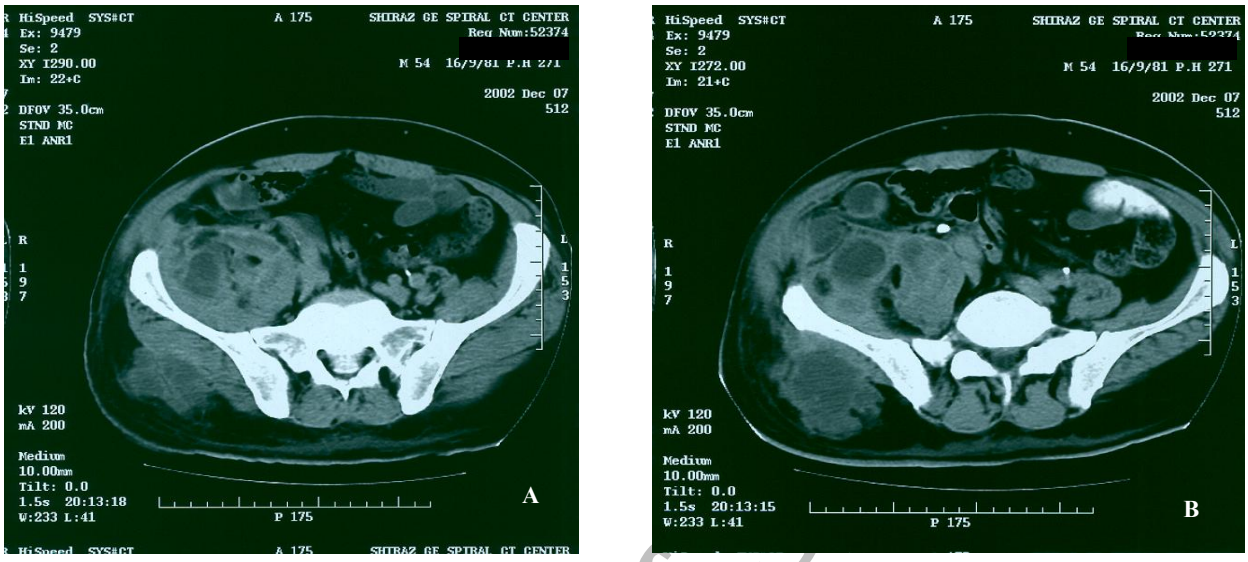

Figure 1 (A and B). Axial spiral contrast enhanced CT-scan at the level of right iliac fossa reveals a combined cystic and solid mass with lobulated borders in the right iliac fossa extending to the right buttock.

A 64-year-old man from southern Iran presented with a one-year history of intermittent right sided lower extremity pain and limping. Since three months prior to his referral, pain had mainly focused in his right hip and buttock. His family and past medical histories were unremarkable. The patient was not receiving any medications. On physical examination, he was conscious and had no difficulty with speech, vision, or respiration. His vital signs were within normal limits. Cardiopulmonary examination was unremarkable and there was no evidence of lymphadenopathy or organomegaly. However, a large nontender mass was found in his right buttock. The patient had two pelvic X-rays, which were reported normal. Abdominopelvic ultrasonography and computed tomography (CT) revealed a dumble-shaped mass with solid and cystic components. The mass involved the right iliac fossa and buttock (Figure 1). Preoperative complete blood count, liver and renal function tests, chest X-ray, and erythrocyte sedimentation rate were all normal.

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Accepted for publication: 18 September 2007

What is Your Diagnosis?

See pages 85 – 86 for the diagnosis.

Photoclinic Diagnosis: **Primary Adenocarcinoma of the Appendix Presenting as a Buttock Mass**

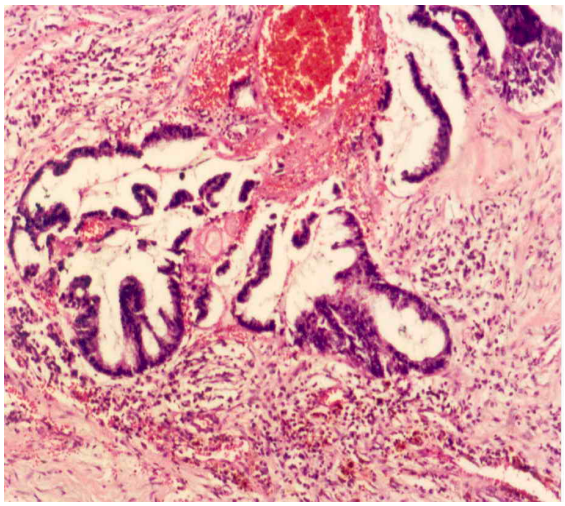

Figure 2 A. Appendix with submucosal lymphoid aggregates, a prominent active germinal center, and muscular layer. Mucosa contains elongated branched mucosal glands, lined by single layer of mucus- secreting epithelium, i.e., villous adenoma (H&E ×4).

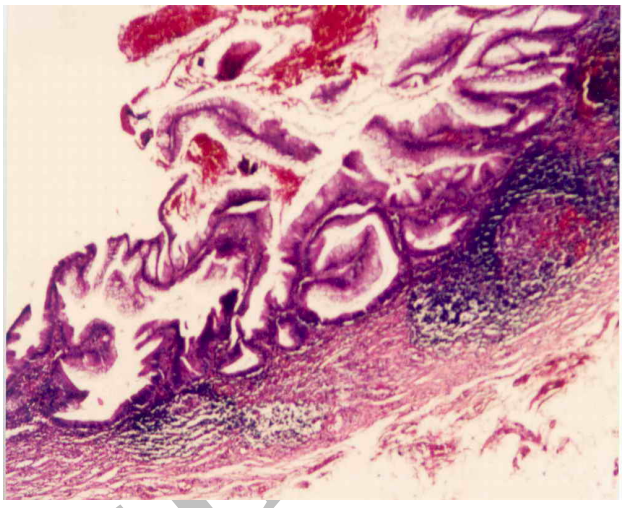

Figure 2 B. Adenocarcinomatous component of the tumor with ma1ignant acini and extracellular mucin invading the stroma (H&E ×40).

The patient underwent exploration with a hockey-stick incision on the right iliac fossa. A cavitating mass was found which was full of gelatinous material. During dissection of the mass, a very tall appendix was found with normal base and body but tumoral tip with attachment to the mass. Partial debulking of the tumor and appendectomy were performed. The histologic diagnosis was mucinous adenocarcinoma of the appendiceal origin with extension to the adjacent tissues (Figure 2). The patient received post-operative chemoradiation therapy, but only one month after completion of the treatment, CT showed a new mass at the previous site. He underwent reoperation. The tumor was totally resected, and right hemicolectomy was performed. The patient has had regular follow-up every three months for 18 months. In the last visit, he was free of disease.

Primary malignant tumor of the appendix is a rare tumor which is difficult to diagnose before surgery because of its frequent unusual presentations. Adenocarcinoma of the appendix is the most common perforating carcinoma of the entire gastrointestinal tract.[1] McCusker et al.[2] reported on 1645 patients with primary malignancy of the appendix. They defined five histologic types, namely mucinous adenocarcinoma, colonic type carcinoma, signet ring cell adenocarcinoma, malignant carcinoid, and goblet cell carcinoid. The age-adjusted incidence of the appendiceal malignancies was 0.12 per 100,000 persons per year. Patients with malignant carcinoid are significantly younger than other patients. Mucinous and signet ring cell adenocarcinoma are more extensive at presentation but goblet cell carcinoids are more limited to the colon. The best overall survival is found in patients with malignant carcinoid and the worse survival is in those patients with signet ring cell adenocarcinoma. Gehrig et al.[3] described five women who presented with symptoms and signs of ovarian tumor but their final diagnosis was appendiceal cancer. They concluded that primary malignant tumor of the appendix should be considered in differential diagnosis of a pelvic mass and the appendix should be evaluated in all patients who undergo operation for an ovarian tumor. Kabbani et al.[4] reported that 70% of patients with mucinous adenocarcinoma presented with pseudomyxoma peritonei, but most patients with nonmucinous carcinoma were operated with the diagnosis of appendicitis. Genetic alterations in appendiceal carcinomas differ from colorectal carcinomas and consist of frequent K-ras mutation but rare alterations of p53. Although most studies showed that mucinous adenocarcinoma of the appendix is an extensive tumor at presentation, as

far as we know, there is no other report in the literature like our patient who presented with a right buttock mass.

References

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4 Kabbani W, Houlihan PS, Luthra R, Hamilton SR, Rashid A. Mucinous and nonmucinous appendiceal adenocarcinomas: different clinicopathological features but similar genetic alterations. *Mod Pathol*. 2002; **15:** 599 – 605.