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MEDI-SEARCH

C/o. Dr. Hedgewar Rugnalaya, Near Gajanan Maharaj Mandir,
Garkheda Parisar, Aurangabad-431 005.
Ph. : 2331195, 2321239. Fax : (0240)339866
e-mail : bamrs@agd2.dot.net.in

Dr. Megha Wani
M.D.(Radio.)

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Dr. Abhishek Parlikar
D.C.H DNB (Ped.)

From the editors' desk

Warm wishes to the readers of Medisearch. As Indians and probably a lot of people world over are ready to tune their bodies to Yoga, in accordance with the celebration of the world yoga day, let us also tune in to whats new in the field of general and Cardiothoracic surgery.

Time and again abdominal lumps have proved to be a matter of surprise for the surgeons; the abdomen, being therefore very aptly named as the pandora's box, poses a lot of diagnostic difficulties. One such case, that our surgeons came across at Dr. Hedgewar hospital, is sure to give you clinical & academic feast. The importance of pre- operative diagnosis is very rightly emphasized in this article.

Cardiothoracic surgery is yet another challenging branch which becomes even more difficult when we come across cardiac patients suffering from major non-cardiac ailments that require immediate surgical correction. Next in line is a similar case where the didactic multidisciplinary efforts and a commendable team work of superspecialists salvaged the life of a cardiac patient.

With every edition of Medisearch we have a Quiz where you can test your clinical accumen. We are more than happy to share the fact that we have been blessed with a lot of readers who attempt to answer the quiz and many of them prove the supremacy of their diagnostic skills. With this edition too we have a clinico-radiological quiz in pediatrics that will surely prove to be a good brain teaser.

-Editors.



IN THIS ISSUE

1 Mucocele of Appendix: a rare case senario

Narendra Kulkarni, Satyajeet
Pathrikar, Praneet Mankar
Dept of Surgery

2 Combined modified radical mastectomy with Aortic valve replacements: a team approach

Vinod Shetkar, Satyajit Pathrikar, Renu Chauhan, Makrand Naik,
Milind Kulkarni, Akshay Nathani and Sachin Ghuge.
Dept of Surgery, Cardiology and Cardiothoracic surgery.
Dr Hedgewar Rugnalaya, Aurngabad, Maharashtra.

3 Quiz

Mucocele of Appendix: a rare case senario

*Narendra Kulkarni, Satyajeet Pathrikar, Praneet Mankar,
Dept of Surgery*

Mucocele is a rare pathology of the appendix, characterized by a cystic dilation of the lumen with mucus stasis. The incidence ranges between 0.2% and 0.3% of all appendectomies, with a higher frequency in females (4:1) and in the people more than 50 years¹. The mucoceles are caused by mucinous cystadenomas and cystadenocarcinomas. In the latter case, a possible rupture of the mucocele, either spontaneous or accidental, may result in the clinical condition of pseudomyxoma peritonei, a spread of malignant cells throughout the peritoneal cavity in the form of multiple mucinous deposits^{2,3}. A correct diagnosis may help to avoid iatrogenic rupture during surgery. We describe a case of a giant appendiceal mucocele and present the diagnostic aspects, surgical options and prognosis of this disease.

CASE REPORT

A 62 year old man presented with diffuse abdominal pain and discomfort. No urinary symptoms or any recent changes in his bowel habits was reported. General examination revealed stable vitals (Pulse rate of 78/min, regular; blood pressure of 140/80 mm Hg taken in right arm supine position) Abdominal examination revealed tenderness in right iliac fossa, no guarding. There was no organomegaly and the rest of the systemic examination was normal.

An abdominal ultrasound revealed a space occupying soft tissue lesion in right iliac fossa measuring 11.9 x 4.9 cm. C.T scan of the abdomen showed a well defined fluid density in lower abdomen s/o mucocele of appendix, the lesion was closely abutting the caecum on right side.

Surgical exploration revealed a giant mucocele of appendix Fig 2.; there were no locally enlarged lymphnodes. No other tumors were noted in the abdomen or pelvis. Appendicectomy was performed. Fig 2.



Fig 1. : The mucocele seen during Laparotomy



Fig 2.: Excised mucocele.

The mass, which measured 20 cm in length and 6 cm in diameter, showed a thin fibrous wall and viscous hemorrhagic mucoid content without any mucosal lesion. The pathological diagnosis was mucinous cyst adenoma. Patient's post operative course was unremarkable; he was discharged on the fifth post-operative day. At 2 month's post operative followup, the patient was asymptomatic.

DISCUSSION

The term mucocele is often used as a general descriptive term for dilatation of the appendiceal lumen by mucinous secretions. Four subgroups of the disease can be identified, according to the characteristics of the epithelium:

- (1) simple or retention mucoceles resulting from obstruction of the appendiceal outflow, usually by a fecalith, and characterized by normal epithelium and mild luminal dilatation up to 1 cm.
- (2) Mucoceles with hyperplastic epithelium where luminal dilatation is also mild; these constitute 5-25% of mucoceles.
- (3) Benign mucoceles, the most common form, accounting for 63-84% of cases. These exhibit mostly epithelial villous adenomatous changes with some degree of epithelial atypia, and are characterized by marked distention of the lumen up to 6 cm. Our patient belongs to this group.
- (4) Malignant mucinous cystadenocarcinomas, representing 11-20% of cases. They are distinguished from the previous group by their glandular stromal invasion and/or presence of epithelial cells in the peritoneal implants. The luminal distention is usually severe.^{1,4}

Mucinous cystadenomas and cystadenocarcinomas are often referred to as neoplastic mucoceles. In cystadenomas, which are the most common form, the luminal dilatation reaches up to 6 cm and is associated with appendiceal perforation in 20% of instances. This results in mucinous spillage into the peri-appendicular area and peritoneal cavity.⁵ Mucinous cystadenocarcinomas, on the other hand, are less common than mucinous cystadenomas and may present with spontaneous rupture in 6% of cases. Macroscopically, they produce mucin-filled cystic dilatation of the appendix indistinguishable from cystadenomas. However, neoplastic cells may penetrate the appendiceal wall and spread beyond the appendix in the form of peritoneal implants. In its fully developed state, the peritoneal cavity becomes distended with adhesive, semi-solid mucin in which neoplastic adenocarcinomatous cells can be found; this condition is termed as pseudomyxoma peritonei. The intraperitoneal spread of this mucin-secreting cancer is identical to that of intra-peritoneal ovarian mucinous cystadenocarcinomas.^{2-5.}

The most common presentation is right lower quadrant pain, similar to an acute appendicitis; a palpable mass can be found in 50% of cases, whereas urinary dysfunction or hematuria is rarely related⁶ Preoperative diagnosis is difficult due to the nonspecific nature of the disease. The lesion may be identified by sonographic which shows a cystic, encapsulated lesion, firmly attached to the cecum, with liquid content and an internal variable echogenicity

Elevated carcino-embryonic antigen levels have been described in neoplastic mucoceles.^{10,11} Computed tomography scan of the abdomen is important in the diagnosis and evaluation of the extent of the disease. A typical computed tomography scan finding of an appendiceal mucocele is a round, low-density, thin-walled, encapsulated mass, communicating with the cecum. related to mucus density. Colonoscopy may show a pathognomonic image, the 'sign of the volcano', i.e., an erythematous, soft mass with a central crater, from which mucus is discharged. Therapy is surgical, but laparoscopic approach is not advised because of the risk of the rupture of soft mass.¹²⁻¹⁵

Appendectomy is used for simple mucocele or for cystadenoma, when the appendiceal base is intact. Caecal resection is indicated for cystadenoma with a large base, and a right hemicolectomy is recommended for cystadenocarcinoma.

Postoperatively, patients with simple or benign neoplastic mucoceles have shown an excellent prognosis with a 5-year survival rates of 91-100%, even in cases with extension of mucus into the extra-appendiceal spaces. In malignant mucoceles, however, the 5-year survival rate is markedly diminished (25%) due to complications of pseudomyxoma peritonei.¹⁸

In conclusion, patients with mucoceles can present with various symptoms, but may also be asymptomatic. Preoperative diagnosis is rare, but it is possible using appropriate tests. These uncommon and potentially lethal entities are usually surgically curable, if diagnosed in an early phase.

Therefore, pre-operative recognition with a carefully planned resection to remove the mass is required.

Literature has evidences of many large mucocele, but in our case the mucocele, happened to be the largest we have ever evidenced . Even the Indian literature shows no record of such a large mucocele. Literature suggest right hemicolectomy as a treatment for mucocele of appendix, but we decided to do minimal surgical intervention by doing simple appendectomy and managed the case successfully.

REFERENCES

- 1 Aho AJ, Heinonen R, Lauren P. Benign and malignant mucocoele of the appendix. *Acta Chir Scand*1973; 139: 392-400
- 2 Crawford J. Tumors of the appendix. In: Cotran R, Kumar V, Robbins S, eds. *Pathologic basis of disease*. Philadelphia Saunders1994; 824-825
- 3 Landen S, Bertrand C, Maddern GJ, Herman D, Pourbaix A, de Neve A, Schmitz A. Appendiceal mucoceles and pseudomyxoma peritonei. *Sur Gynecol Obstet*1992;175: 401-404
- 4 Higa E, Rosai J, Pizzimbono CA, Wise L. Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendix: A re-evaluation of appendiceal mucocele. *Cancer*1973; 32: 1525-1541
- 5 Gibbs NM. Mucinous cystadenoma and cystadenocarcinoma of the vermiform appendix with particular reference to mucocele and pseudomyxoma peritonei. *J Clin Pathol*1973; 26:413-421
- 6 Merran S. Tumeur muco-secretante de l'appendice (mucocele appendiculaire). *Presse Med*1997; 26: 933
- 7 Peek DF, Beets GL. Pseudomyxoma peritonei in the pleural cavity: report of a case. *Dis Colon Rectum*1999; 42: 113-116
- 8 Takahashi S, Furukawa T, Ueda J. Case report: mucocele of the tip of the appendix. *Clin Radiol*1998; 53: 149-150
- 9 Stevens KJ, Dunn K, Balfour T. Pseudomyxoma extraperitonei: a lethal complication of mucinous adenocarcinoma of the appendix. *Am J Gastroenterol*1997; 92: 1920-1922
- 10 Dachman AH, Linchtenstein JE, Friedman AC. Mucocele of the appendix and pseudomyxoma peritonei. *Am J Roentgenol* 1985; 144: 923-929
- 11 Soweid AM, Clarkston WK, Andrus CH, Janney CG. Diagnosis and management of appendiceal mucoceles. *Dig Dis* 1998; 16: 183-186
- 12 Kim SH, Lim HK, Lee WJ, Lim JH, Byun JY. Mucocele of the appendix: ultrasonographic and CT findings. *Abdom Imaging* 1998; 23: 292-296
- 13 Zissin R, Gayer G, Kotos E, Apter S, Peri M, Sharipo-Feinberg M. Imaging of mucocoele of the appendix with emphasis on the CT findings: a report of 10 cases. *Clin Radiol*1999; 54: 826-832
- 14 Isaacs KL, Warshauer DM. Mucocele of the appendix: computer tomographic, endoscopic, and pathologic correlation. *Am J Gastroenterol*1992; 87: 787-789
- 15 Hamilton DL, Stormont JM. The volcano sign of appendiceal mucocele. *Gastrointest Endosc* 1989; 35: 453-456
- 16 Aho AJ, Heinonen R, Lauren P. Benign and malignant mucocele of the appendix: Histological types and prognosis. *Acta Chir Scand*1973; 139: 392-400

Combined Modified Radical Mastectomy with Aortic Valve Replacements: A Team Approach

Vinod Shetkar, Satyajit Pathrikar, Renu Chauhan, Makrand Naik,
Milind Kulkarni, Akshay Nathani and Sachin Ghuge.
Dept of Surgery, Cardiology and Cardiothoracic surgery.
Dr Hedgewar Rugnalaya, Aurangabad, Maharashtra.

Introduction:

Aortic stenosis (AS) is narrowing of the aortic valve, obstructing blood flow from the left ventricle to the ascending aorta during systole. Bicuspid aortic valve (BAV) is the commonest cause of isolated aortic stenosis below the age of 70 years¹⁻⁵. This condition often co exists with other morbidities, in which scenario a multidisciplinary approach is required for prioritizing the therapy options. In this article we discuss the management of a 38 yr old woman with symptomatic AS (due to a BAV) coexisting with an invasive breast carcinoma. Dynamic decision making and a coherent execution of therapeutic efforts helped us in treating this patient.

The Case:

A middle aged premenopausal patient with a bicuspid Aortic Valve and severe Aortic Stenosis (Valve area 0.8 sq.cm and mean gradient of 65mmHg) was diagnosed to have a lump in her left breast during the perioperative evaluation for valve replacement surgery. The lump (8cm x 6cm in size) was present in the left upper quadrant with palpable lymph nodes in the left axilla. Fine needle aspiration cytology revealed invasive ductal carcinoma. As per the oncologist's opinion the lump needed to be addressed early as a life saving procedure.

Discussion between the Cardiologist, the Oncosurgeon and the Anesthesiologist, regarding the pros & cons of isolated procedure, ended with the decision of combining both the procedures in one setting. Consequently, patient underwent *Auchincloss modified radical mastectomy* with axillary dissection followed by *aortic valve replacement with no 21 St. Jude Standard Valve* in one setting.

Operative details:

Auchincloss modified radical mastectomy-

Under general anesthesia with all invasive monitoring in supine position with shoulder support and left axilla elevation, the modified radical mastectomy with axillary dissection was performed first. Through an elliptical incision, the entire breast tissue along with the tumour including a 2 cm margin of skin and the nipple-areola complex were removed. This was followed by axillary dissection with clearance of all the axillary fat pad and lymph nodes up to the stage III. Pectoralis muscles were retracted upwards, entire axillary vein was cleared preserving the neural structures. Complete hemostasis was achieved, Romovac suction drains were placed, one underneath the skin flap and into the axilla below the pectoralis muscles and incision closed in layers without any tension. Skin was closed with surgical staples.

Aortic valve replacement -

After this a midline sternotomy was done and pericardium opened & hitched. Heparin given 3mg per kg body weight and ACT kept above 400s through the procedure. Patient was put on *cardiopulmonary bypass* and myocardium was protected with direct and *retrograde coronary cold cardioplegia*. *Standard oblique aortomy* was done 2cm above the origin of right coronary artery and aortic valve assessed. The bicuspid valve, with fused and calcified left and right coronary cusps, was completely excised and replaced with a no. 21 St. Jude standard mechanical heart valve. Aortomy was closed in two layers. Heart deaired and the patient was slowly rewarmed and taken off bypass with minimal inotropic support. All throughout the the procedure she was hemodynamically stable.

On post-op day one she was extubated, with the removal of mediastinal drains done on day two and she was started on Low Molecular Weight Heparin and warfarin (INR was kept between 2 to 3). Skin flap part of romovac drain was removed on post operative day 7 and the axillary part on post-operative day 24. Patient was discharged on the 10th postoperative day and we have a regular follow up of her till date. All skin sutures were removed between 15th through 24th postoperative day. All wounds healed well and there were no areas of flap necrosis or dehiscence at any of the incision sites.

6 weeks postoperatively she was started on chemotherapy. Radiotherapy was also given as an adjuvant for local disease control. It is 8 months past the procedure now and she is doing well.

Discussion:

Aortic stenosis (AS) is the obstruction of blood flow across the aortic valve. Patients become symptomatic gradually after a latent period of 10-20 years. Common etiologies of AS are degenerative, rheumatic and congenital (unicuspid or bicuspid)⁵⁻¹⁰. Bicuspid valve is the commonest cause of isolated severe AS in patients less than 70 years of age, with an overall prevalence of 0.5 to 2%. Once symptomatic, mortality is 25% at 1 year and 50% at 2 years. In asymptomatic patients with moderate to severe stenosis, incidence of sudden cardiac death is 4% per year. The classic triad of symptoms is angina, syncope and exertional dyspnoea¹⁻⁵. Diagnosis is based on two dimensional echocardiography which is the imaging modality of choice to diagnose and estimate the severity of aortic stenosis and to localize the level of obstruction. Normal aortic valve area is 2.5-3.5 sq.cm. Patients are classified as mild (1.5-2.5), moderate (1-1.5), severe (<1) and critical (less than 0.8sq.cm) depending upon the narrowing of this area (as shown in the respective brackets). The only definitive treatment for aortic stenosis is aortic valve replacement (AVR)¹¹⁻¹⁴.

AS is a major risk factor for peri-operative cardiac events in patients undergoing noncardiac surgery. Combined procedures or other major non-cardiac procedures in patient with severe AS are becoming more common due to ageing population. Most common associations are with malignancies of other organs or with emergency surgeries^{11-13,15,16}. We, at Dr. Hedgewar hospital, faced a similar situation where the patient had breast carcinoma as a co-morbidity associated with AS.

Both, invasive ductal carcinoma and severe AS, are life threatening and warrant early therapy. Only dilemma lies in prioritizing the ailment to be treated first. The disadvantage of doing the AVR first followed by radical mastectomy is the chances of excessive bleeding as the patient is on anticoagulation therapy post AVT. If mastectomy is done first, we are likely to face the problems of valve dehiscence and paravalvular leak due to immunocompromised status. Also, two major surgeries in two different settings, increases the overall cost and morbidity due to prolonged hospitalisation in addition to the individual risk of mortality during each procedure.

Hence we decided to perform both the procedures in one setting. With one time anaesthesia and hospitalisation we were able to do both the surgical procedures with an excellent result.

References:

1. Friedman T., Mani A., Elefteriades J.A., : Bicuspid aortic valve: Clinical approach and scientific review of a common clinical entity. *Exper Review cardiovascular Ther.* 6 2008: 235-248
2. Ward C., Clinical significance of bicuspid aortic valve. *Heart* 83 2000:81-85
3. Tezemos N, Therrien J, Yip J.; Outcome in Adults with bicuspid aortic valve, *JAMA* 300 2008:1317-1325
4. Fedak P. W., Verma S, David T. E., Leask R. L., Weisel R. D., Butany J., : Clinical and pathophysiological implications of a bicuspid aortic valve. *Circulation.* 106 2002: 900-904.
5. Warnes C. A., Williams R.G., Bashore T. M., : ACC/AHA 2008 Guidelines for the management of adults with congenital heart disease. *J Am Coll Cardiology* 52 2008 e1-e121
6. Pellika P. A., Sarano M. E., Nishimura R. A., ; Outcome of 622 adults with asymptomatic hemodynamically significant aortic stenosis during prolonged follow up. *Circulation.* 111 2005:3290-3295.
7. Rosenhek R., Binder T., Porenta G., : Predictors of outcome in severe asymptomatic aortic stenosis. *N Eng J Med.* 343 2000;611-617.
8. Russo C.F., Mazzetti S., Garatti A. : Aortic complication after bicuspid aortic valve replacement ; longterm results *Ann Thoracic Surg.* 74 2002 : s1773-s1776. Discussion s1792-9.
9. Rutherford SD, Braunwald E. : Aortic Stenosis ; *Heart diseases: A textbook of cardiovascular medicine*, 4th ed. Philadelphia: WB Saunders.
10. Otto CM.: Timing of aortic valve surgery, *Heart.* 2000;84:211-21
11. Davidson's Principles and practise of medicine(21st ed.) Edinburgh: Churchill Livingstone/Elsevier. 2010.
12. Arpino G. Bardou VJ, Clark GM, Elledge RM, (2004).; Infiltrating lobular carcinoma of the breast, tumour characteristics and clinical outcome. *Breast Cancetr research.* 6(3): R149-156.
13. Dixon JM, Anderson TJ, Page DL, Lee d, Duffy SW, : Infiltrating lobular carcinoma of breast. *Histopathology* 1982 (6): 149-161.
14. National comprehensive cancer Network, breast cancer. V1 2014. National comprehensive network clinical practice guidelines in oncology. 2013.
15. Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine
16. Hurst's the Heart, 13th Edition

Now its Quiz time !

A 36 hour old term male newborn, presented with irritability, abdominal fullness, refusal to feed since 6-8 hours. The baby had not passed stools since birth. There was no vomiting. Have a look at the x ray below and enlist at least three differentials for this clinico-radiological case scenario.



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Address for Correspondence :
Editor - MEDI-SEARCH
C/o, Dr. Hedgewar Rugnalaya,
Garkheda Parisar, Aurangabad-431 005.
Phone - (0240) 2331195, 2321239
e-mail - abhishekarlikar@gmail.com