

PRESENTATION OF BREAST DISEASES -3 (NIPPLE PROBLEMS)

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Abstract: Nipple problems are the problems/diseases related to nipple areola complex. Complete absence of the breast and nipple is rare. Accessory nipple is the presence of extra nipple present in the mammary line. Nipple Discharge is a complex diagnostic challenge for clinicians. All patients with nipples discharge need careful assessment with history and examination. The majority 95% will be benign in origin. The nipple discharge can be classified according to its color, cellularity and biology. The discharge has to be true, spontaneous, persistent and non lactational to be significant.

Key words: Duct ectasia, Intraductal papilloma, Galactorrhoea

NIPPLE PROBLEMS

These are the problems/diseases related to nipple areola complex.

CONGENITAL

- Absence.
- Supernumerary.
- Bifid.
- Inversion.

ACQUIRED

- Inversion.
- Plasma cell mastitis.
- Mammary duct fistula.
- Duct ectasia.

- Retroareolar carcinoma.
- Paget's disease.
- Amazia, Amastia



Accessory nipple
Courtesy Prof. Asif Zafar

Complete absence of the breast and nipple is rare. It may be associated with failure of development of pectoralis muscles (Poland's syndrome).

SUPERNUMERY NIPPLE

It is the presence of extra nipple present in the mammary line. It is neither a common abnormality nor has any significance. It doesn't need any treatment either.

The breasts are modified sweat glands and

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are derived from ectodermal mammary ridge, which extends from axilla to groin region.



Supernumery Nipple

In many animals breasts develop along the whole length of this ridge, but in human only a pair develop in the pectoral region. Sometimes supernumery or only nipple may be seen along this line¹⁰.

BIFID NIPPLE

The nipple is divided into two parts usually from the middle congenitally. The nipple may be inverted or normal. It is usually associated with recurrent infection. It is rare but well defined entity. No treatment is required. The patient is reassured.



Bifid Nipple



Bilateral inversion of nipples due to congenital chest deformity. Courtesy Faisal Bilal Lodhi, FCPS

INVERSION OF NIPPLE

Inversion of nipple is a condition which is characterized by inward displacement of the tip of nipple. It leads to appearance of a slit like opening and disappearance of almost whole of the nipple.

CONGENITAL

Congenital failure to protrude nipple is also called inversion. Incidence of inversion is quite high. About 20% of females present with this problem. They should be reassured.

These patients mostly need no treatment.



Nipple retraction with Peau-de-Orange (Cancer Breast) Courtesy Sajid Shiekh, FCPS

Sometime they present with the difficulty is suckling by the baby. Nipple sheath is applied to the nipple for helping the baby to suck easily. It also helps to get the nipple everted.

ACQUIRED

Acquired inversion of nipple is mostly due to duct ectasia, plasma cell mastitis or any other inflammatory lesion. Retro areolar carcinoma may also lead to inversion of nipple.

NIPPLE RETRACTION

It is the inward displacement of whole/part of nipple from its base in the direction of disease process. It is usually due to malignancy. The lesion may be present in the breast tissue at any part. The retraction is due to involvement of connective tissue Bands (ligaments of Astley Cooper).

It is acquired problem which could be due to duct ectasia, carcinoma or surgical problem. All these problems are discussed at length, at some other part of the book.

MAMMARY DUCT FISTULA

Mammary duct fistula is an aftermaths of periductal mastitis. It presents as a discharging sinus around the areolar margin.



Mammary Duct Fistula

DUCT ECTASIA

This is a common disease process. Its etiology is not known. It is not clear whether ectasia or inflammation is the primary problem. No one is clear which starts first. Sloughing of ducts initiates a process of periductal mastitis. Clinical features follow the pathological changes. Duct ectasia presents as nipple discharge nipple retraction or a palpable mass.

The terminal lactiferous ducts enlarge in size and then dilate grossly. The epithelial lining disintegrate and plugs the dilated lactiferous ducts.

Multicolored fluid from the nipple is discharged. Periductal chemical mastitis starts. It is infiltrated with plasma cells and Foreign Body Giant cells.

A hard indurated mass with overlying inflammation may appear at the areolar margin.

This inflammation may resolve automatically or change into periareolar abscess. If it is not treated adequately it may lead to mammary duct fistula. The repeated episodes of periductal mastitis leads to fibrosis along ducts which shrink and may lead to inversion of nipple.

It is commonly seen in postmenopausal women but it may occur in premenopausal women. It is often bilateral and multiple abscesses and fistulae may be seen.

Treatment is surgery. Duct excision is performed to exclude malignancy if the discharge is blood stained.

PAGET'S DISEASE OF BREAST

Paget's disease is macroscopic superficial skin manifestation of underlying carcinoma of breast. Usually it presents as a unilateral nipple ulceration. Sometimes whole of nipple is eroded and it completely disappears. The cancer of breast becomes evident sooner or later if it is not treated in time.



Paget's disease of nipple.

It is a breast lesion which presents as a chronic eczematoid eruption of nipple areola complex. It was first described by Sir James Paget in 1874.

It is seen in approximately 02% of patients. The specific demonstration of Carcino-Embryonic Anti-gen (CEA) within the paget's cell facilitates in diagnosis and its differentiation from malignant melanoma lesions. It occurs in old age group of patients. Very rarely it may even present as early as 20 years of age.

Clinically Paget's disease of nipple presents with an itching and vesicular eruption involving the nipple areola complex.

Paget's disease of breast is a type of ductal carcinoma which extends from the ducts to involve the skin of nipple and areola. The

malignant invasion of the nipple presents as eczematous change of unilateral nipple and areola. It may present as an encrusted and scaly tumor present at nipple and areola.

The involved skin is ulcerated, fissured and oozing. It may have surrounding area of inflammatory hyperaemia and edema. Advanced cases show total ulceration or even loss of nipple and areola. There may be superimposed infection and suppurative necrosis. Its symptoms include tenderness, itching, burning and intermittent nipple discharge.

The underlying mass or lump is rarely palpable.

The epidermis is involved by the malignant cells and these are called. "Paget's cells". Their origin and nature is unknown. The intraductal lesion is often multifocal and ducts throughout breast may be dilated due to obstruction of central collecting ducts at the ampulla.

Paget's cells are large, anaplastic and hyperchromatic ovoid cells surrounded by a clear zone or halo. These have pale cytoplasm. It is possibly due to ballooning degeneration. The anaplastic tumor cells lie singly or in clusters within the epidermis.

NIPPLE DISCHARGE

Nipple Discharge is a complex diagnostic challenge for clinicians. All patients with nipples discharge need careful assessment with history and examination. The majority 95% will be benign in origin. Salient features in history include;

- Unilateral/bilateral
- Single/multiduct
- Colour - particularly the presence of

blood

- Spontaneous or only with expression
- Presence of an associated lump

A variety of diseases, such as intraductal papillomas, mammary duct ectasia, breast cancer, pituitary adenomas, breast abscesses and infections can manifest as nipple discharge¹.

Nipple discharge disorders is a field in which there has been both increasing awareness on part of patients and advancements in management.

The nipple discharge can be classified according to its color, cellularity and biology. The discharge has to be true, spontaneous, persistent and non lactational to be significant³.



Lactating breast showing milky and blood stained discharge.

PHYSIOLOGICAL DISCHARGE

It usually presents with staining of the under garments or bed linen. The discharge is multiductal and maybe white, yellow, green, brown, or black.

- Normal milk discharge during pregnancy and lactation.
- Normal discharge at abnormal time.

Lactation

This is the most common cause of nipple discharge and needs no treatment. This is physiological and milk discharge usually stops after cessation of breast feeding. In parous women milk can be expressed out even after two years of cessation of lactation.

Some times pregnant patient can have blood stained discharge which settles on its own. Occasionally neonates may show milk discharge from their nipple if luteal or placental hormones get entry into fetal circulation again it needs no treatment.

PATHOLOGICAL DISCHARGE

It is usually spontaneous, unilateral and single duct involvement is seen. The discharge may be serous, serosanguis or blood stained. It is caused by various diseases such as given following.

PATHOLOGICAL

- Intraductal papilloma
- Duct ectazia
- Carcinoma
- Fibrocystic disease
- Trauma
- Infection



Blood stained nipple discharge
Courtesy Sajid Shiekh, FCPS

SEROUS DISCHARGE

- Duct ectasia.
- Epithelial hyperplasia.
- Green discharge.
- Blood stained discharge.
- Grumous discharge.
- Pus discharge.

GALACTORRHOEA

It is bilateral copious, milky discharge from multiple ducts. Prolactin levels should be checked and if raised > 100 m/u/liter. The cause is due to a pituitary prolactinoma. Other secondary causes of galactorrhoea include some medicine and ectopic hormone secretion.

It is the discharge of milk from nipple which is unrelated to breast feeding or lactation. It can be physiological after cessation of feeding but continuous mechanical stimulus to nipple promotes milk discharge.

DRUG RELATED

Drugs which reduce production of prolactin

- Dopamine.
- Tricyclic antidepressants.
- Methyl dopa.
- Cimetidine.
- Benzodiazepine.

Drugs which block dopamine receptors.

- Phenothiazide.
- Metachlopramide.
- Hexachlopramide.
- Haloperidol.
- Oestrogen.
- Digitalis.

SPONTANEOUS GALACTORRHEA

Pituitary adenoma producing prolactin
Broncho-genic carcinoma.

- Cushing's syndrome
- Hypothyroidism

BLOOD STAINED NIPPLE DISCHARGE

It causes high degree of anxiety in women because of fear of breast cancer. Most frequently it is benign. It is commonly caused by intra ductal papilloma, duct ectasia and less frequently by carcinoma breast².

Discharge from nipple can occur from single or more lactiferous ducts. The discharge may be serous, serosanguis, blood stained, yellowish or black. Nipple discharge accounts for 5% of all referrals to breast clinics in UK. Cause of nipple discharge is benign and in 10-20% of cases, malignancy may be detected in 80% to 90% cases.

Discharge may be from peri areolar breast skin. Which may occur due to different skin diseases like eczema, psoriasis or chancre. Peri areolar discharge can also be seen in Paget's disease and duct ectasia.

TRIPLE ASSESSMENT

Correct diagnosis requires triple assessment. Appropriate planning of management is done after assessment of the patient and understanding of disease process.

CLINICAL EXAMINATION

A detailed clinical evaluation by triple assessment is invaluable to determine the pathophysiology and assess risk of malignancy¹.

Both breasts and axillae are examined. Nipple is cleaned and breast is squeezed to see nature and site of discharge and number of ducts involved.

Hemocult Test is performed to confirm presence of blood in the discharge. Galactography, ultrasound examination, exfoliative cytology are useful².

A large number of false negative results make these tests less productive and histopathological tests are required to confirm the diagnosis².

SERUM PROLACTIN LEVEL

When all other investigations are normal and patient has got amenorrhoea along with visual disturbances, then raised serum prolactin level may suggest pituitary adenoma. Sometimes serum prolactin may rise in bronchogenic carcinoma.

IMMUNOLOGICAL TESTS

Modern immunological tests can be performed on cytology smears where occurrence of high levels of carcino-embryonic antigen (CEA) could indicate latent malignancy³.

IMAGING

SONOMAMMOGRAPHY

High resolution ultrasound is helpful in visualizing intra ductal abnormality and are becoming a good complimentary approach if not an alternative to traditional radiology techniques³.

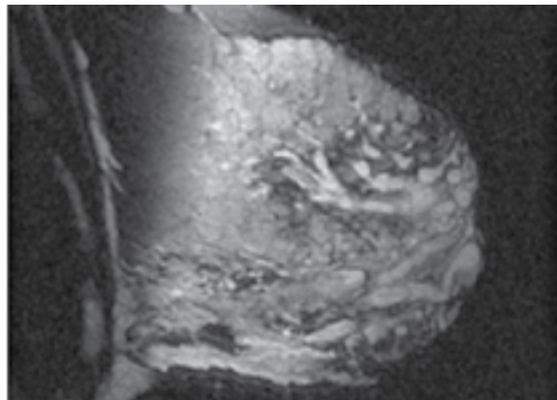
Ultrasound is more sensitive than galactography in cancer diagnosis. It also permits guided biopsy and pre operative localization of impalpable ductal lesions. It can be used as a complimentary diagnostic tool⁶.

MAMMOGRAPHY

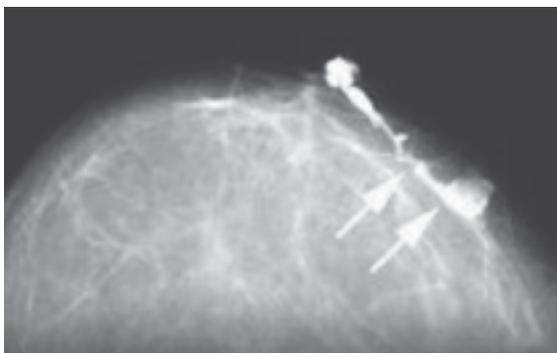
This investigation is again not very helpful in finding out the cause of nipple discharge but some time hidden impalpable mass may be detected by mammography.

Absence of palpable mass or mammographic abnormality gives a false sense of security

causing delay in diagnosis².



Ductography of lactating breast.



Serous discharge from nipple.
(Ductography)



Serosanguis discharge from nipple
(Ductography)

DUCTOGRAM (GALACTOGRAPHY)

Galactography is the state of art approach to investigate patients with nipple discharge disorder and this examination can demonstrate the size, location and extent of an intra ductal abnormality³.

It may be helpful in diagnosing intra duct papilloma.

MR galactography has been shown to be of diagnostic value but not as informative as radio-galactography³.

FIBROPTIC DUCTOSCOPY

Early success of image guided excision of papilloma and duct endoscopy promise a significant improvement in diagnostic accuracy through minimally invasive procedure.

The most sophisticated investigation method which can be used therapeutically as well is fibroptic ductoscopy of the concerned duct in a breast.

It is a new technique and is expensive. It is a fascinating and promising approach for visualizing the intra ductal lumina³.

Mammary ductoscopy for pathological nipple discharge is a safe, effective procedure that offers advantages of high lesion localization rate and intra operative guidance, therefore, negating the need for pre-operative ductogram. Lesions deep within the ductal system can be identified and removed which could have been missed by blind duct excision⁵.

It can be helpful to pick up cells for cytology. Its role is yet controversial.

CYTOLOGICAL EXAMINATION

Discharge is examined under the microscope to rule out malignancy but false negative reports may confuse the management plan. Ductal lavages in combination with cytology provide promising results².

Cytology smears of discharge material have helped to classify the cellular material, providing information about normality, atypia and malignancy and also about papillary formation³.

MANAGEMENT

Objectives of treatment are to;

1. To rule out malignancy.
2. To reassure the patient.
3. To offer definitive treatment of the causative factors

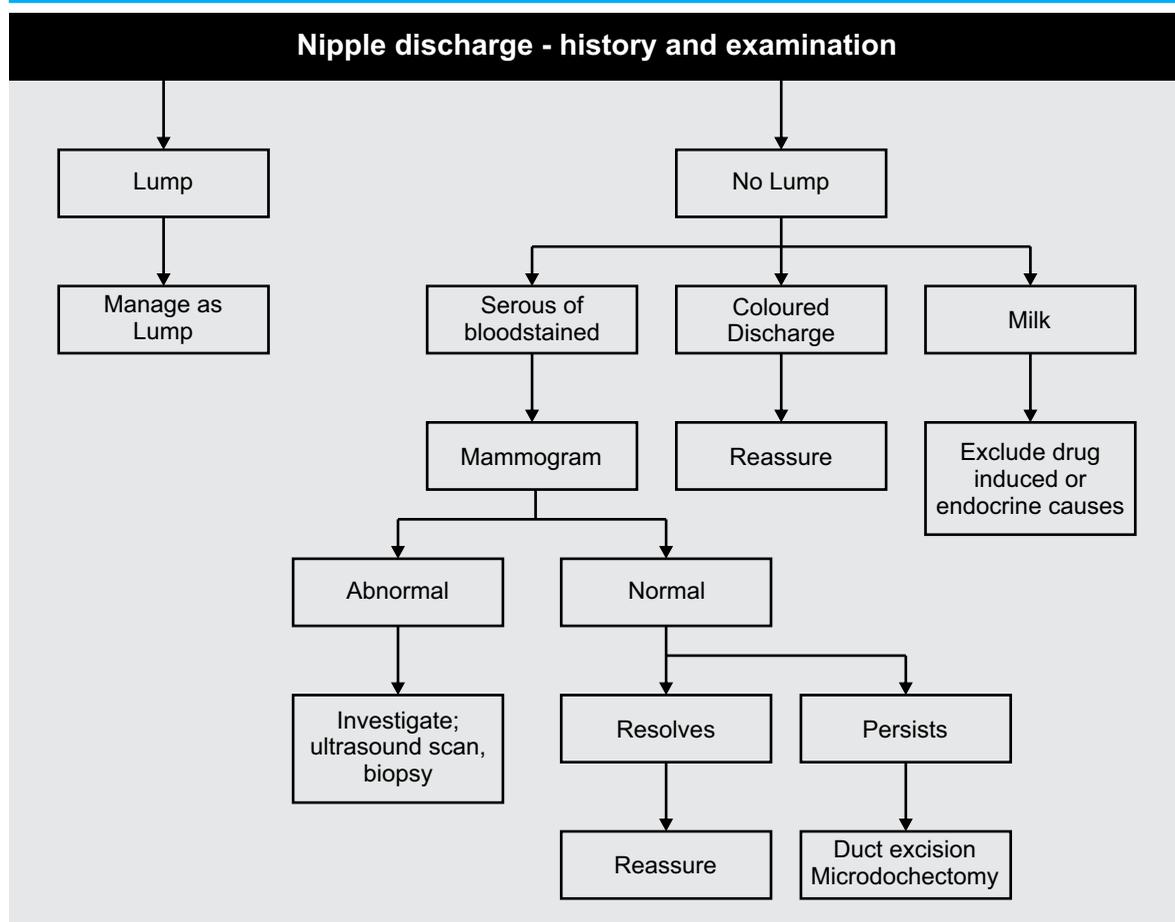
SURGERY

Indications of surgery;

- a. Single duct discharge
 - i. Blood stained discharge
 - ii. Mammography shows mass
 - iii. Troublesome discharge
 - iv. A new symptom in sixty years old patient
- B. Multiple ducts discharge
 - i. Troublesome.
 - ii. Persistent.
 - iii. Copious.

The patients with nipple discharge should be clearly differentiated from those who require surgery and who don't need it. Surgically significant nipple discharges are watery, serous (yellow), serosanguis or blood stained³.

Women presenting with pathologic nipple discharge require duct exploration regardless of cytologic or radiologic findings.



When discharge is the result of local extensiveness of disease and intra ductal spread, it may preclude breast conservation in more than 60% of cases⁴.

Nipple discharge in males is very rare but once it is there, it should be regarded with suspicion. There are high chances of underlying malignancy. Surgical excision of the involved ductal system from which the discharge emanate. It is the only reliable procedure in establishing a certain diagnosis and controlling the blood stained discharge². Histologically it is an extension of intra ductal carcinoma or even infiltrating ductal carcinoma. 30-40% of these patients have

metastasis at the time of surgery and prognosis is less favorable than simple intra ductal carcinoma which is diagnosed at earlier stage.

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