Mujer 36 años técnico de Rx que consulta por atrofia lineal en ambos muslos
Mujer de 49 años que consulta por > de volumen en brazos
Lipoatropia semicircular: Mecanismos patogénicos

- Microtraumatismos

- Campos electromagnéticos generados por equipos informáticos y mucho cableado

- Descarga electrostática por diferencia de potencial entre el cuerpo y el borde de la mesa

- Humedad por debajo del 30 por ciento

- Estimulación del Macrófago, liberación de citoquinas: TNF alfa lesivo para el adipocito
Osakidetza
H. CRUCES
GURUTZETAKO OSPITALEA
CRUCES-BIZKAIA

Consultas Externas

MEDICINA INTERNA CONSULTAS

Historia / Nº Historia: 80588
EIk / CIP: 39254
Gizarte Seguranza / Nº Seguridad Social: 480106993483
Jalex Data / Fecha Nacimientoa: 11/09/1967
Zerbiltza / Servicio: Profesional / Profesional
M. INTERNA CONSULTA: ALVAREZ BLANCO, ALFONSO

FECHA DE CITA: 21/01/2010 FECHA INFORME: 25/01/2010

INFORME:
MOTIVO DE CONSULTA: Atrofa lineal en ambos muslos.

ANTECEDENTES PERSONALES:
- Mastopatia fibroquistica.
- Espondilitis (herencia familiar no compresiva).
- Episodio de sinovitis en cadera derecha hace un año.

ANTECEDENTES LABORALES: Técnico de rayos.

ENFERMEDAD ACTUAL: Desde hace un mes nota atrofia lineal en tercio medio de ambos miembros inferiores.

EXPLORACION GENERAL: Dentro de la normalidad. Destaca lívido reticularis y lipoatrofia semicircular en ambos miembros inferiores.

ANALITICA Y EXPLORACIONES:
- Hematometra, perfil general, T4, TSH y autoinmunidad: todo en rango de la normalidad.

RMN DE PARTES BLANDAS: mínima irregularidad del contorno cutáneo medio distal del compartimento graso anterior del muslo.

J. DIAGNOSTICO:
- LIPOTROFIA SEMICIRCULAR.

RECOMENDACIONES:
- Se remite a Salud Laboral para evaluación de su puesto de trabajo, pues este cuadro está descrito en relación con campos electromagnéticos, disminución de humedad... etc., y en este caso está en relación cronológica con su actividad.

Med Clin (Barc) 2002;119(10): 390-5. web articulo 45.508
Med Clin (Barc) 2008;130(6): 213-5. web articulo 206.272
http://www10.gencat.net/treball_scst
Mujer de 49 años que consulta por > de volumen en brazos

- Consulta 24/12/2008 por > volumen de brazos desde hace 6 meses
- Antecedentes personales
  - Técnico fotográfico
  - 2 embarazos normales
  - Déficit de factor Von Willebrand
- Enfermedad Actual:
  - Etilismo activo desde hace más > seis meses
  - Desde hace doce meses lipomas en brazo y aumento progresivo del perímetro de ambos brazos con parestesias en manos
• Analitica:
  – Glucosa , Hb. Glicosilada normal
  – GOT::167,GPT:50, GGT: 1060. PCR:<0.3.
  – VIH:negativo
  – ECO hepatico: esteatosis grasa
  – TAC abdominal: suprarenales normales.
    Esteatosis . No HP
APARTAMENTOS TUTELADOS EN BARAKALDO
¡ÚLTIMOS A LA VENTA!
TU PISO EN PROPIEDAD
Venta Directa del Promotor
RMN TORAX, PELVIS Y EXTREMIDADES SUPERIORES

- DEPOSITO DE GRASA SUBCUTANEA EN RAIZ DE BRAZOS > 4 CM

- AUSENCIA DE GRASA SUBCUTANEA EN CUELLO

- EN RESUMEN:
  - DEPOSITO DE GRASA PREFERENCIAL EN HOMBROS Y EXTREMIDADES INFERIORES

CONSULTA ENDOCRINOLOGIA:

LIPODISTROFIA PARCIAL ADQUIRIDA CEFALOTORACICA
Clasificación de las lipodistrofias

Lipodistrofias hereditarias Generalizadas
Lipodistrofia congénita generalizada (síndrome de Berardinelli-Seip)
   - Displasia mandíbulo-acral tipo B
   - Lipodistrofias asociadas a progeria
Parciales
   - Lipodistrofia familiar parcial
   - Forma de Dunnigan
   - Forma de K6bberling
   - Displasia mandíbulo-acral tipo A

Lipodistrofias adquiridas
Generalizadas
   - Lipodistrofia generalizada adquirida (síndrome de Lawrence
Parciales
   - Lipodistrofia parcial adquirida (síndrome de Barraquer-Sirnorzs)
   - Lipodistrofia secundaria a tratamiento antirretroviral (en HIV
Lipodistrofias localizadas
   - Inducidas por fármacos
   - Por presión, por flujos electromagnéticos
   - Secundarias a paniculitis
   - Idiopáticas
Patients with acquired lipodystrophies

(A) Lipodystrophy in a man with human immunodeficiency virus who received antiretroviral therapy containing a protease inhibitor. Note the loss of fat from the face, arms, and legs, increased fat, and buffalo hump.
(B) Acquired partial lipodystrophy in a woman with Barraque syndrome. The loss of subcutaneous fat from the face, neck, thorax, and upper abdomen began in adolescence. Note the subcutaneous fat in the legs.
(C) Acquired generalized lipodystrophy. The generalized loss of subcutaneous adipose tissue began in adolescence. Complex severe insulin resistance, hyperinsulinemia, hypertriglyceridemia, serum high-density lipoprotein cholesterol concentrations.
Patients with congenital generalized lipodystrophies

This panel shows a woman and a man with congenital generalized lipodystrophy types 1 and 2. Type 1 is due to a mutation in acylglycerol-3-phosphate O-acyltransferase 2 gene. Type 2 is mutation in the seipin gene. Both types are characterized by generalized lack of fat, extreme musculature, acanthosis nigricans, axillae, and acromegaloid features. Patients with CGL1 have higher prevalence of intellectual impairment and cardiomyopa than those with CGL1.
## Clinical features of patients with lipodystrophic disorders

<table>
<thead>
<tr>
<th>Feature</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lipodystrophy</td>
</tr>
<tr>
<td>Acanthosis nigricans (associated with severe hyperinsulinemia)</td>
</tr>
<tr>
<td>Muscle hypertrophy, prominent veins</td>
</tr>
<tr>
<td>Hepatomegaly</td>
</tr>
<tr>
<td>Fatty liver</td>
</tr>
<tr>
<td>Cirrhosis</td>
</tr>
<tr>
<td>Hypertrichosis, hirsutism (occasionally)</td>
</tr>
<tr>
<td>Cardiomegaly due to hypertrophic cardiomyopathy (occasionally)</td>
</tr>
<tr>
<td>Mental retardation (occasionally)</td>
</tr>
<tr>
<td>Metabolic abnormalities</td>
</tr>
<tr>
<td>Hyperglycemia, but no ketosis</td>
</tr>
<tr>
<td>Insulin resistance</td>
</tr>
<tr>
<td>Hypertriglyceridemia</td>
</tr>
<tr>
<td>Increased metabolic rate</td>
</tr>
</tbody>
</table>
Boston, December 17, 2009

Thank you for requesting a second opinion for Mrs. a 49 year old woman, from Spain; with remarkable fat deposit in the upper limbs.

The case was analyzed by Dr. David Baldwin, Director and Associate Professor of Endocrinology at Rush University Medical Center.

We asked him the following questions regarding this case:

1. Do you agree with the provided diagnosis? What would be the cause of her disease?
2. What are the differential diagnoses?
3. Is there any relationship between patient’s lipodystrophy and her past history of alcohol abuse?
4. Do you recommend additional studies?
5. What are the therapeutic alternatives? What are the risks and expected benefits of each option?
6. What treatment do you recommend?
7. Prognosis.
8. Do you have any additional recommendation for the patient or her physicians?
9. Could you please provide several references relevant to this case that the patient and treating physicians may find useful and interesting?
10. Do you know any expert in Spain that you could recommend to follow up this patient?

Attached are his answers to these questions.

I hope this information is useful for the management of this patient.
David Baldwin, MD
Director and Associate Professor of Endocrinology at Rush University Medical Center

Rush University Medical Center
Division of Endocrinology
1725 West Harrison Street, Suite 250
Chicago, IL 60612

Tel: 312-942-6163

Profile:
Medical School: Rush Medical College
Internship: Brigham and Women’s Hospital
Residency: Brigham and Women’s Hospital
Fellowship: University of Illinois at Chicago Medical Center
Certification: Internal Medicine

Specialty/ Subspecialties
General Endocrinology and Metabolism
The patient is a 49 year old woman with a history of alcohol excess has developed progressive fat deposits in her upper arms over the past 23 months. Her weight has increased from 42 to 56 KG over the past several years but she quit ETOH in Dec 2008. She takes no meds, she has no signs of cortisol excess, and she has not lost or gained fat in her neck, chest, abdomen but has gained some fat in her legs but not as much as her arms. Her past history has been summarized and reviewed and is not contributory. She is now menopausal and is not hirsute. Nor does she have the cardinal coetaneous clue for insulin resistance namely acanthosis nigricans. She has had some hepatomegaly in the past suspected to be due to ETOH. There is no recent exam of her liver. Her lab studies rule out diabetes or pre diabetes, her lipid studies are ok, she does not appear to have a clue for hypertena. Her autoimmune studies and complement levels are normal. I do not see a urine study but presume that she does have nephritis/nephrosis. Thyroid studies are normal, FSH and LH are elevated due to menopause, she does not have hyperinsulinemia, and serum cortisol is 19 but unclear what time it was drawn. It is likely a normal AM cortisol but would need to repeat at 8 AM after 1 mg dexamethasone PO at bedtime the night before to rule out cortisol excess. Her pictures and history do NOT suggest Cushing's Syndrome however.

Her local consultations have suggested “acquired partial cephalo-thoracic lipodystrophy”, however I do NOT agree with this assessment.

1. Partial lipodystrophies are usually familial but can rarely be acquired. However one expects to see fatty hypertrophy in one body area associated with lipodystrophy in a different body area. The usual familial form (type 2) usually has loss of fat from extremities and abdomen and an increase in fat in the chin and neck. Elevated blood glucose and lipids are typical. Thus she does not fit. The entity “acquired partial cephalo-thoracic lipodystrophy” is also known as Barraque-Simon’s syndrome. It is characterized by loss of fat in the face and upper trunk and by sparing or increased adiposity in the rest of the body. (see up to date article) There are many other features of this syndrome such as nephritis, diabetes, insulin resistance, and complement changes which she does not have. Thus it just does not fit to my eye. I do not see an HIV study, but I will presume that she does not have HIV associated lipodystrophy because this usually causes abdominal
lipohypertrophy and lipoatrophy of the extremities especially during treatment with HAART. Thus for these reasons I do NOT believe that she actually has a lipodystrophic syndrome, or any other endocrine syndrome.

2. My differential diagnosis was as above
3. I can discover NO relationship between her fat accumulation in her arms and her previous history of ETOH in my review of the literature.
4. I do not recommend further studies except as noted above, and their yield should be low.
5. Various treatments have been proposed for lipos dystrophy (TZD, leptin etc) but none have been actually shown to be effective and again I do not believe that she has a lipodystrophy. Plastic surgery either open or liposuction is well described as being successful in patients like her with localized benign idiopathic fat deposition.
6. Plastic surgery as above can be considered and is usually effective.
7. Prognosis for with localized benign idiopathic fat deposition (my expression) is excellent as long as she does not have any metabolic disturbances which I do not believe that she does.
8. Additional recommendations are only as above
9. I have sent the relevant articles to you from Up to Date which are indeed up to date. My further exhaustive review of PUBMED has not found any further literature that I feel is relevant.
10. I am not aware of specialists in Europe, however I do not believe that she needs further expert medical consultation or management except for perhaps a few tests as noted above and perhaps a plastic surgery referral.

Very sincerely, David Baldwin MD

The articles I will mail to you in Boston today. db
Diagnósticos diferenciales.

Patología tiroidea (bocio, carcinomas, y otros)
Quistes cervicales
Tumores benignos y malignos (hemangiómas, lipomixangiómas, etc)
Cuello de búfalo (enfermedad de Cushing, inhibidores de la proteasa
Enfermedades linfoproliferativas, metástasis linfáticas
Algunas formas de distrofia muscular
Enfermedad de Von Recklinghausen
Lipomatosis familiar múltiple
Lipomatosis dolorosa (Síndrome de Dercum)
Neurolipomatosis de Alsberg
Lipomatosis nodular de Krabbe y Bartels
Lipornatosis de Touraine y Renault
Lipoma múltiples secundarios a lesiones intracraneales (Síndrome de Frohlich)
Pseudolipomatosis de Vemeuil y Potaín
Asociaciones propuestas

Alcoholismo crónico
Neuropatía periférica
Anemia con VCM elevado*
Alteraciones hepáticas*
Hiperuricemia*
Dislipemia
Diabetes mellitus
Hipotiroidismo
Acidosis tubular renal
Carcinoma de vías aéreas superiores
Otros carcinomas (pulmón, lengua, Kaposi, entre otras)
Figure 2: "Pseudoathletic appearance" due to fat tissue distribution on thorax, abdomen and proximal limbs.
TRATAMIENTO

• LIPOSUCCION  2.200 CC DE GRASA

• RDT: TERAPIA ENDODERMICA RADIAL
  – MASAJE MAS DRENAJE LINFATICO